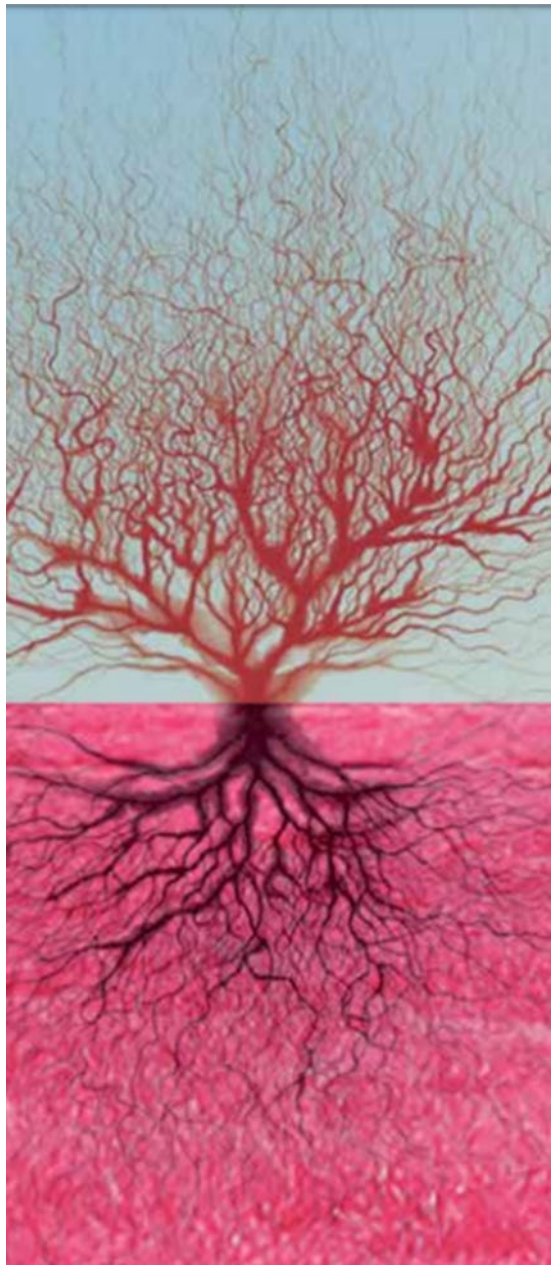
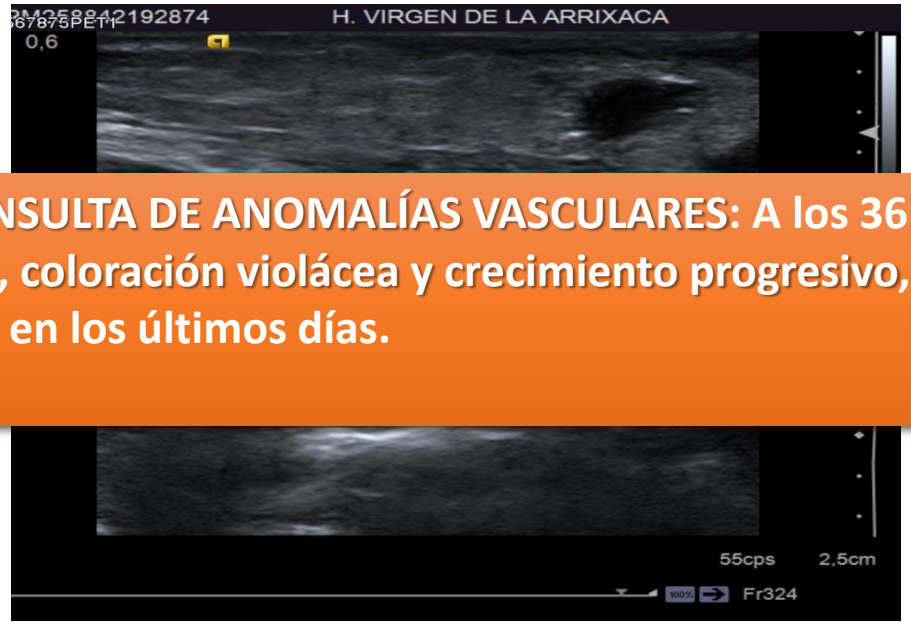


ANOMALÍAS VASCULARES

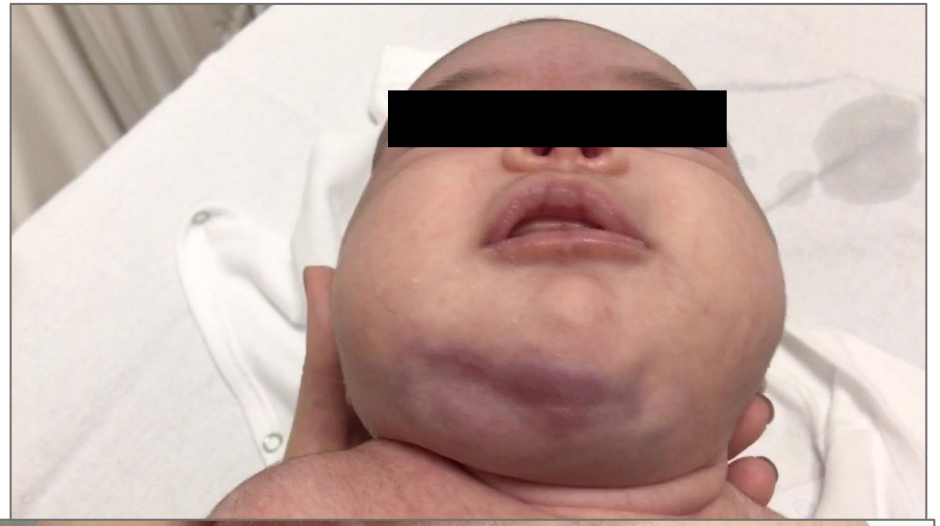
CASO 3

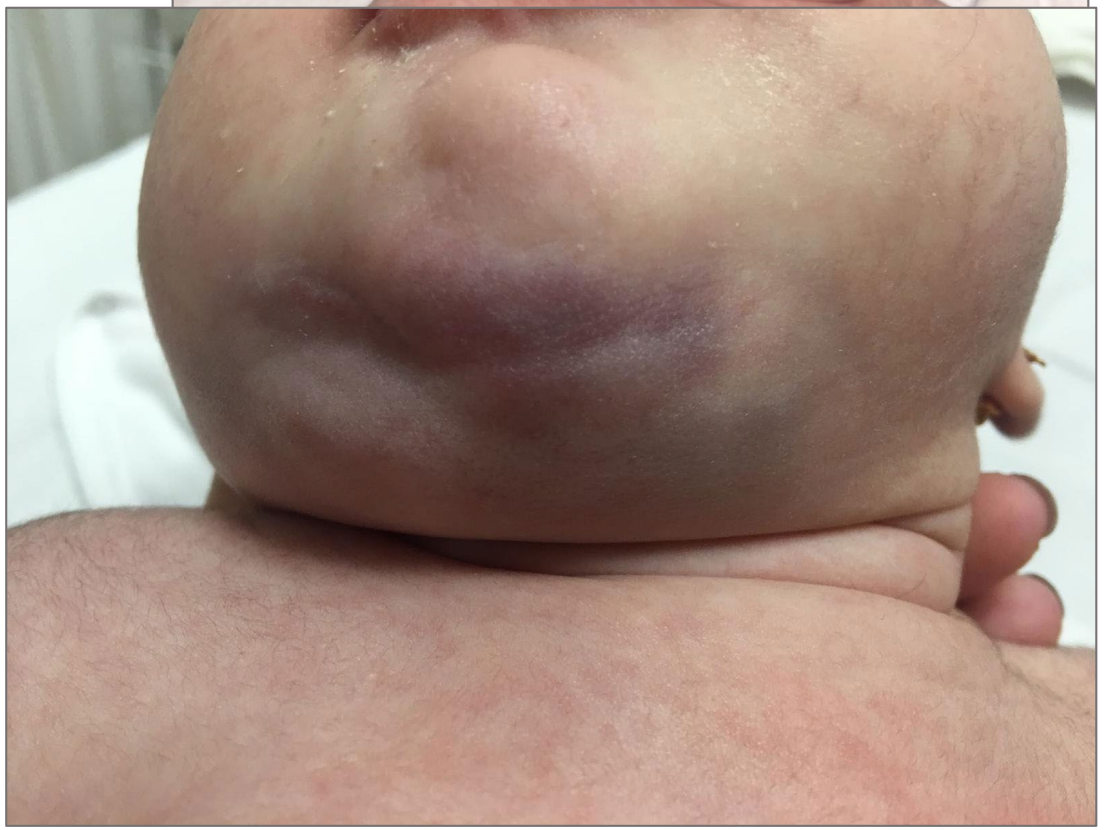
Laura Barona García, María Isabel Ortuño Moreno, Alejandro Salazar Nicolás, Albert Caballero Illanes, Gema Ruiz García, Belén Ferri Ñíguez.

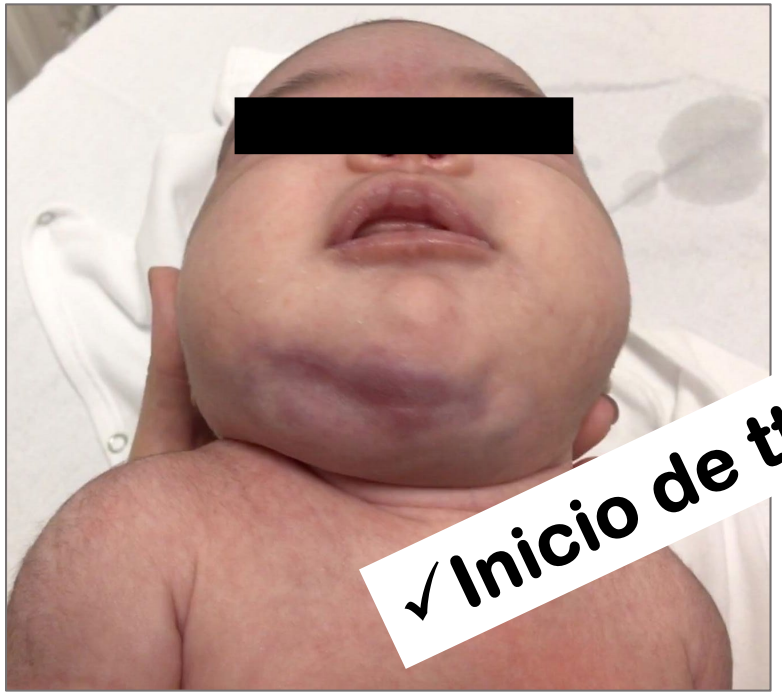
• Recién nacida que a las 24 horas de nacer se detecta asimetría a expensas de tumoración submandibular derecha, inicialmente sin cambio de coloración.
• Ecografía submandibular: En región submentoniana derecha, lesión sólida, hipoecogenica de bordes bien delimitados, y vascularizada al flujo doppler de 7,8x15,6x16,7mm. Todo ello, en relación con tumor de origen vascular, probablemente un **hemangioma infantil en fase proliferativa** como primera posibilidad.

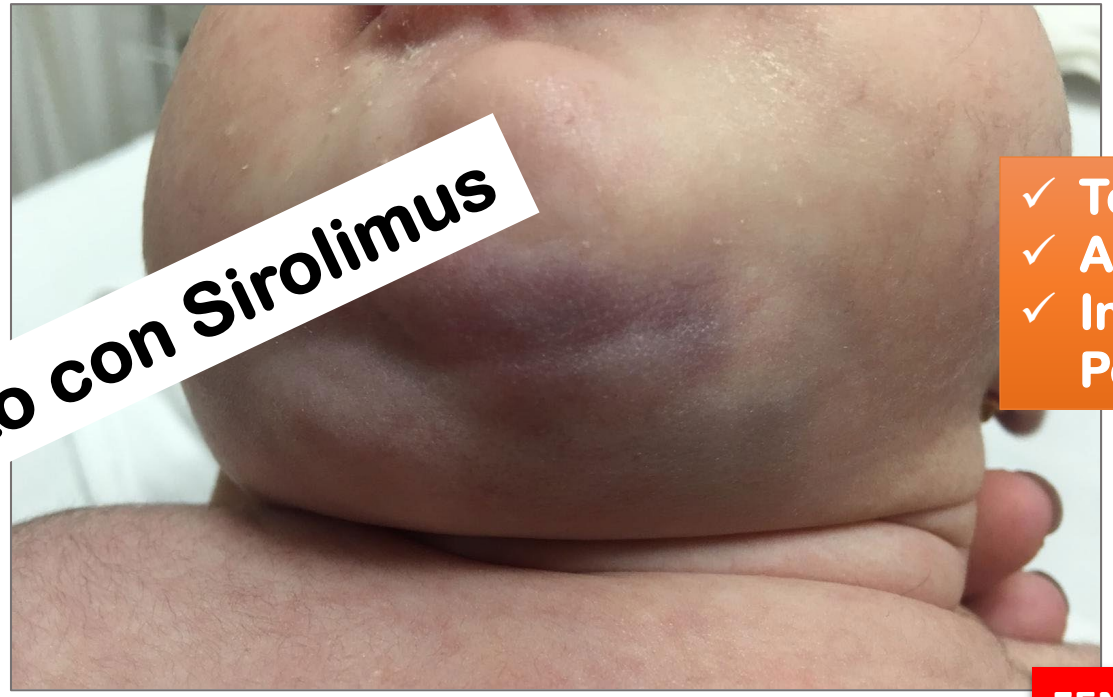


CONSULTA DE ANOMALÍAS VASCULARES: A los 36 días, coloración violácea y crecimiento progresivo, más en los últimos días.





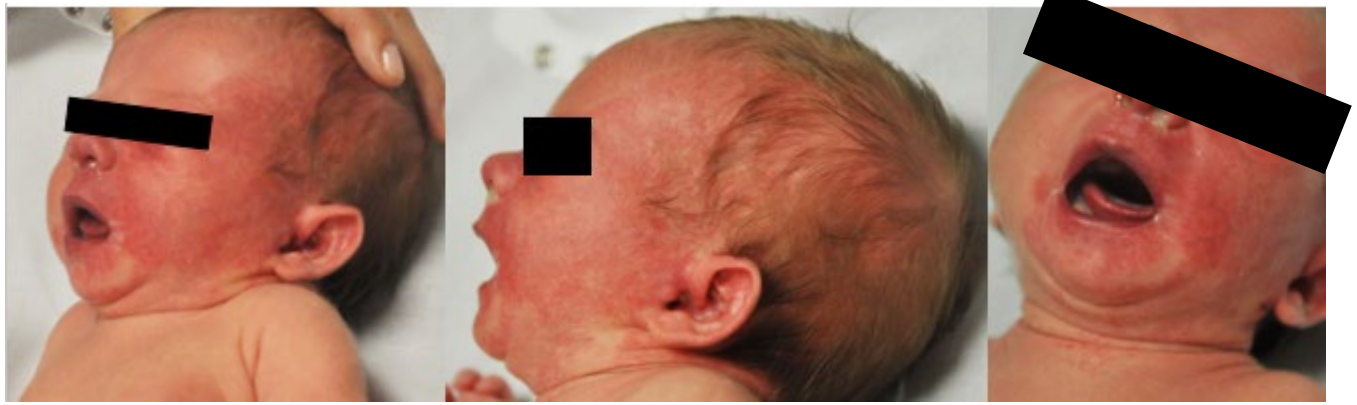




✓Inicio de tto con Sirolimus

- ✓ Toma de biopsia
- ✓ Analítica urgente
- ✓ Ingreso en Cirugía Pediátrica

FENÓMENO DE KASABACH- MERRIT

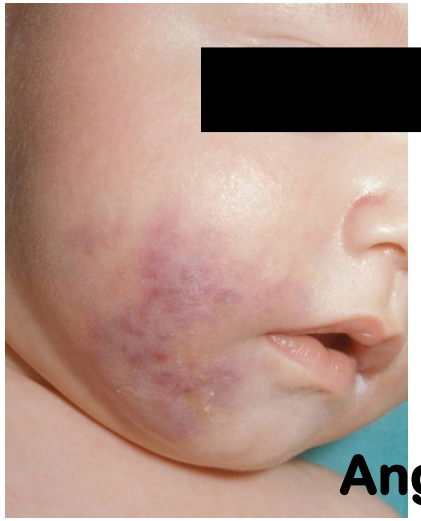


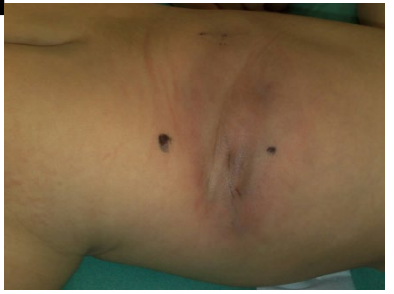
Hemangioma Infantil / Congénito

propanolol

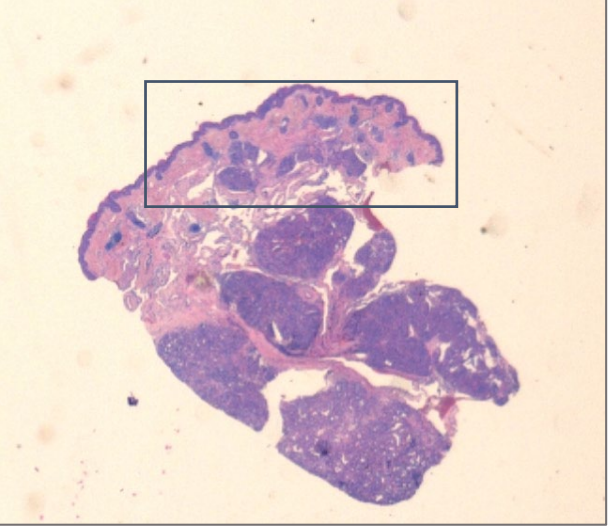
vs

Sirolimus

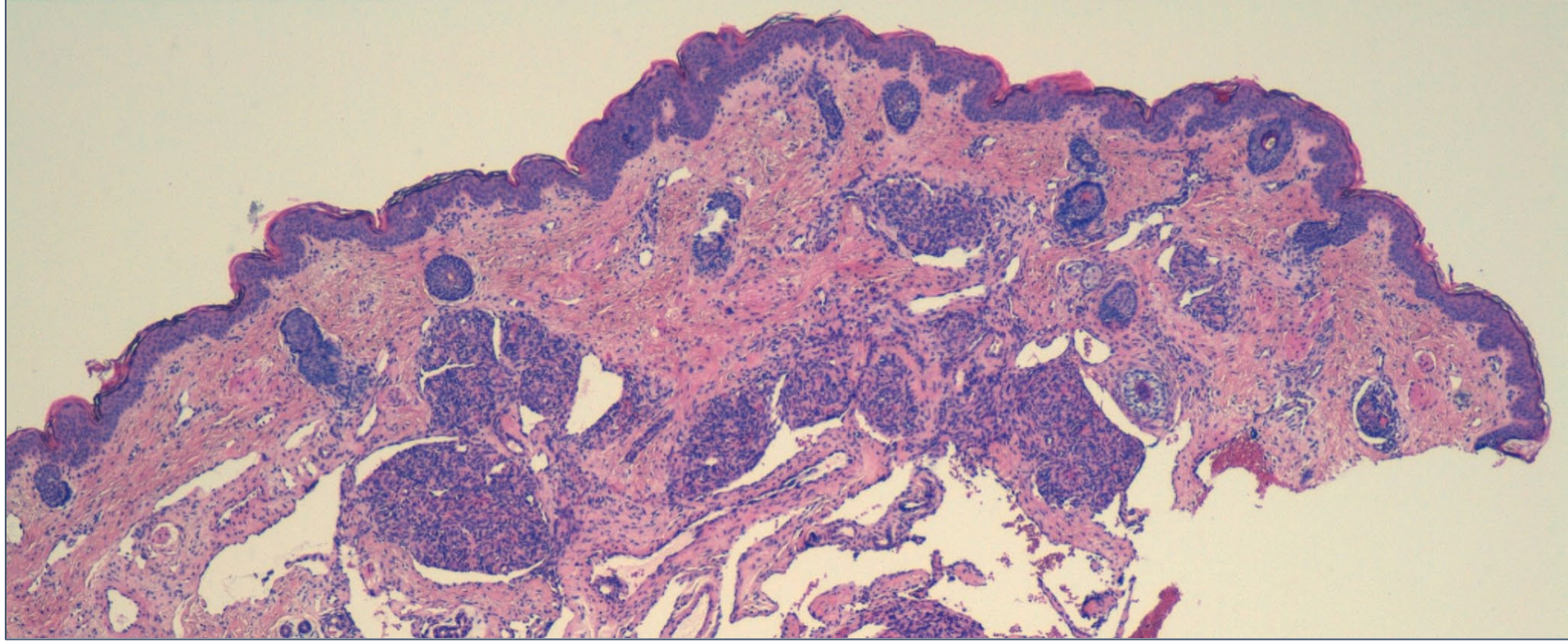


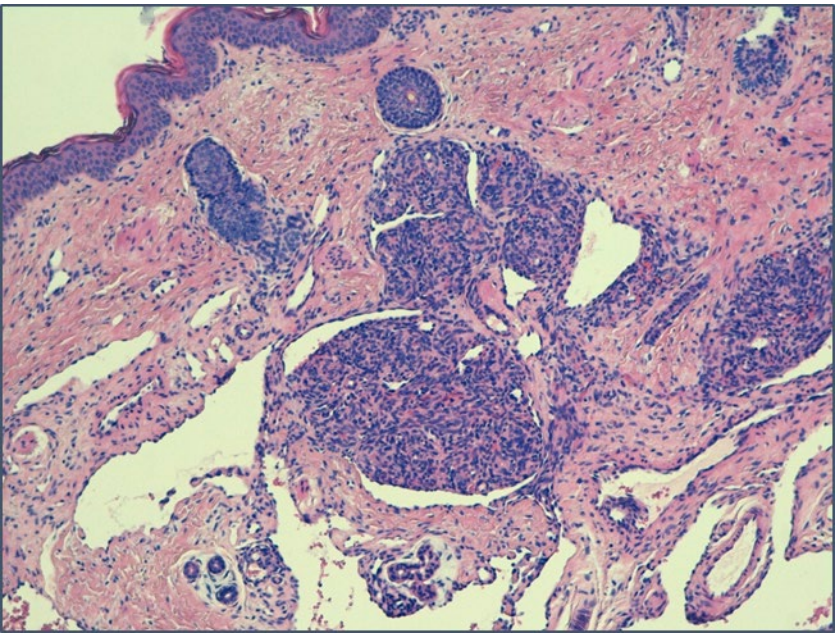


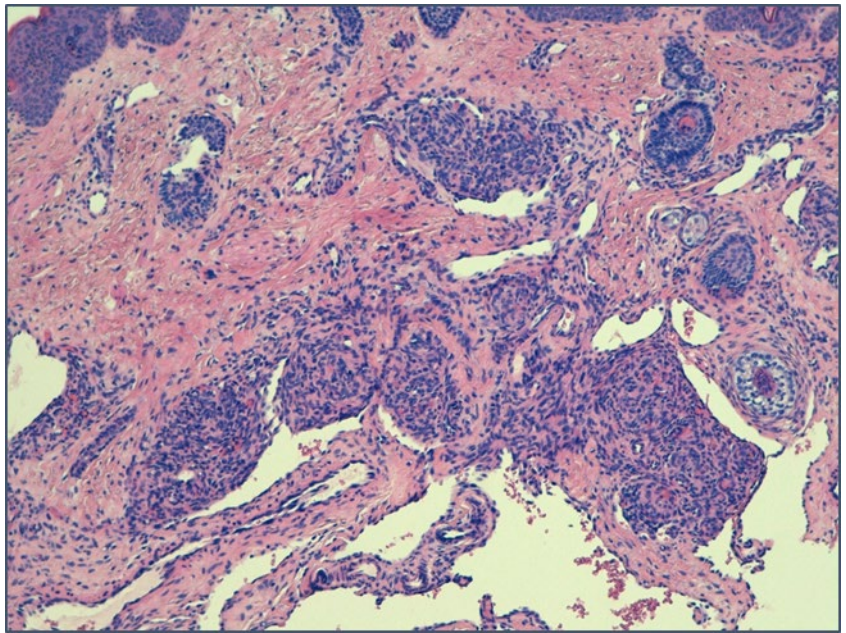
Angioma Tufted / KHE

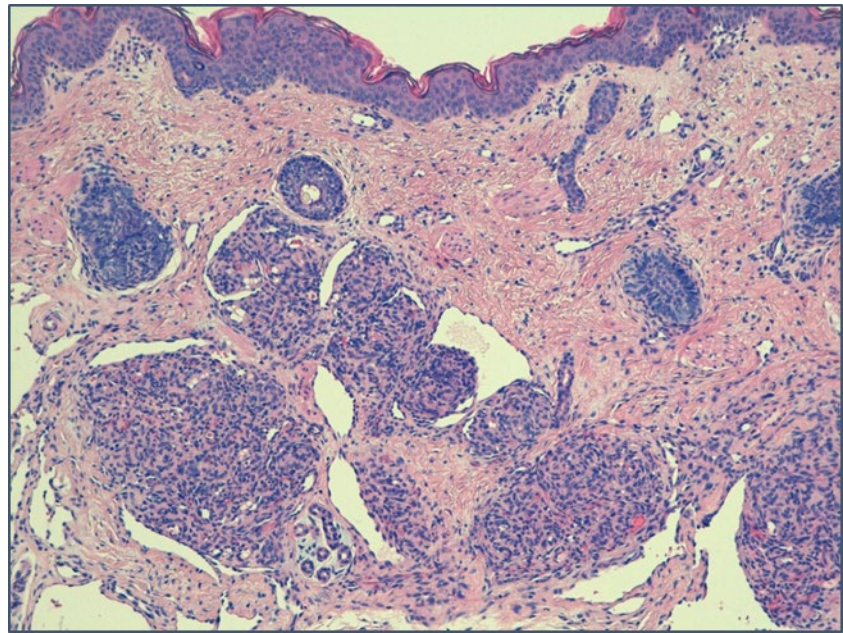


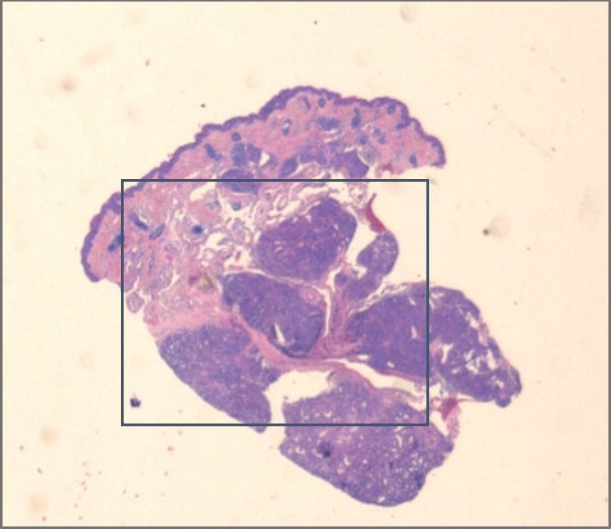
<24 horas: H-E + IHQ

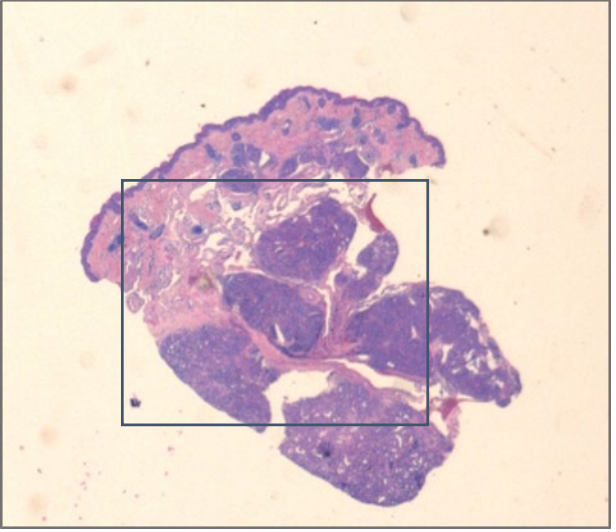


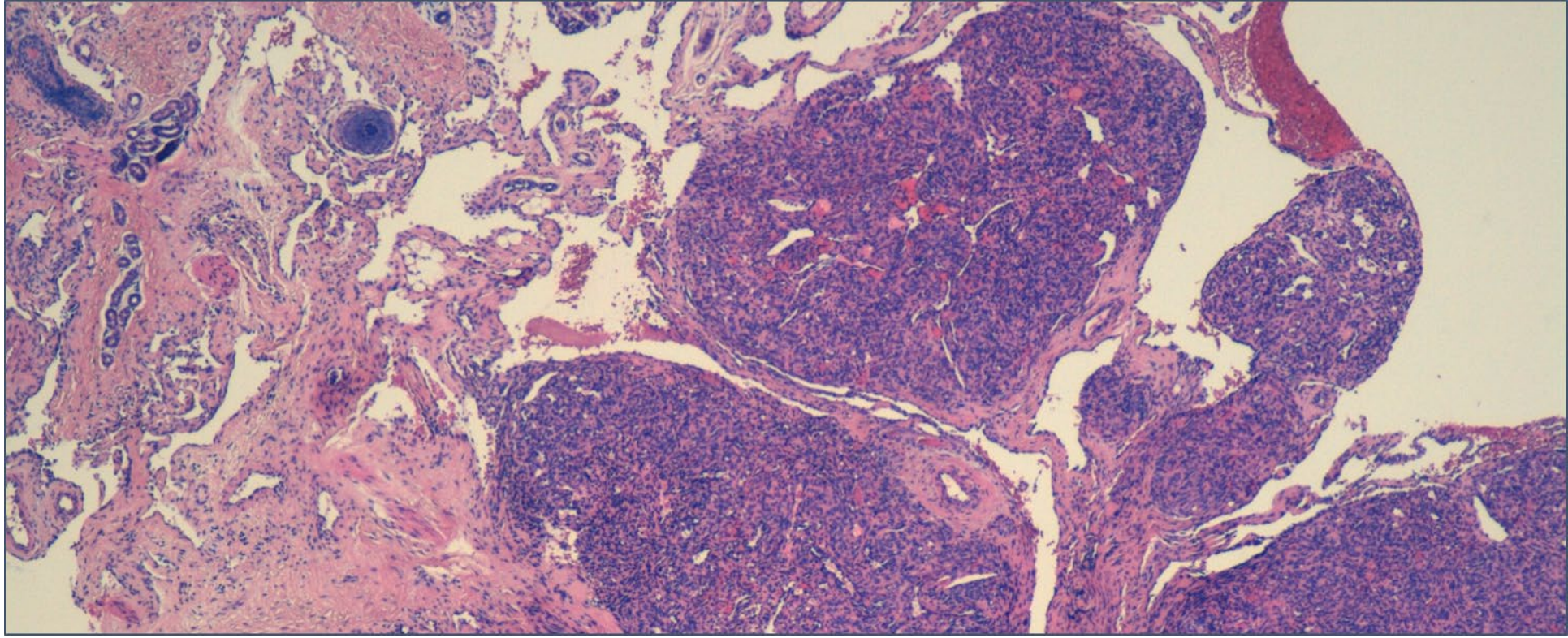


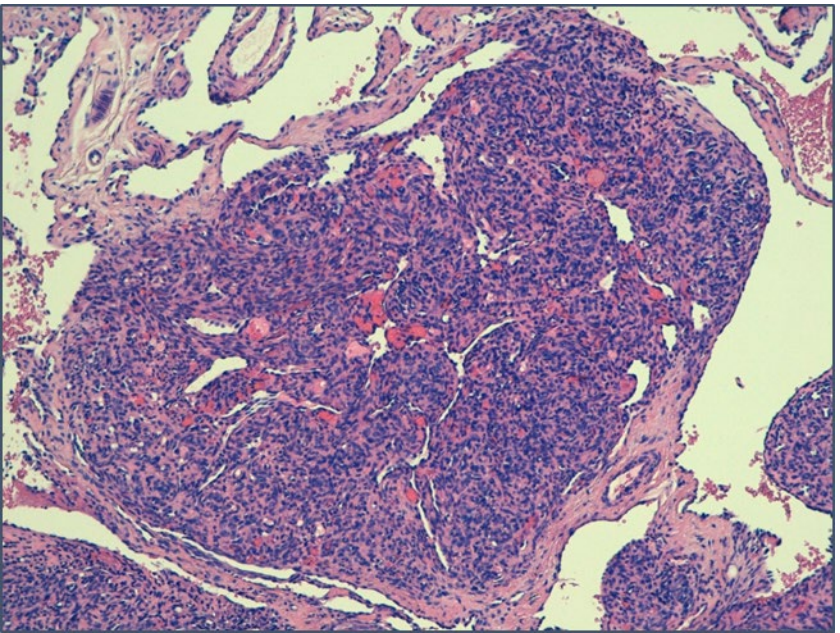


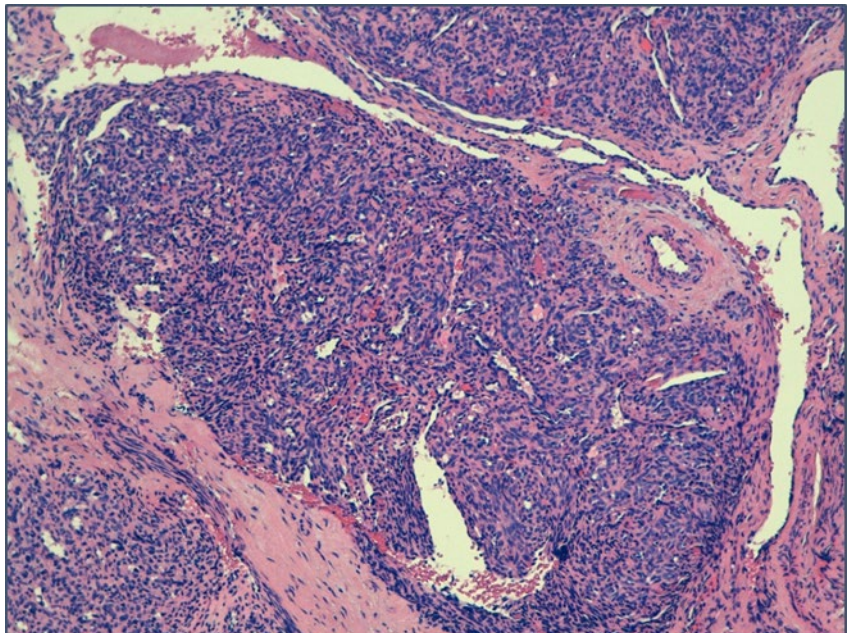


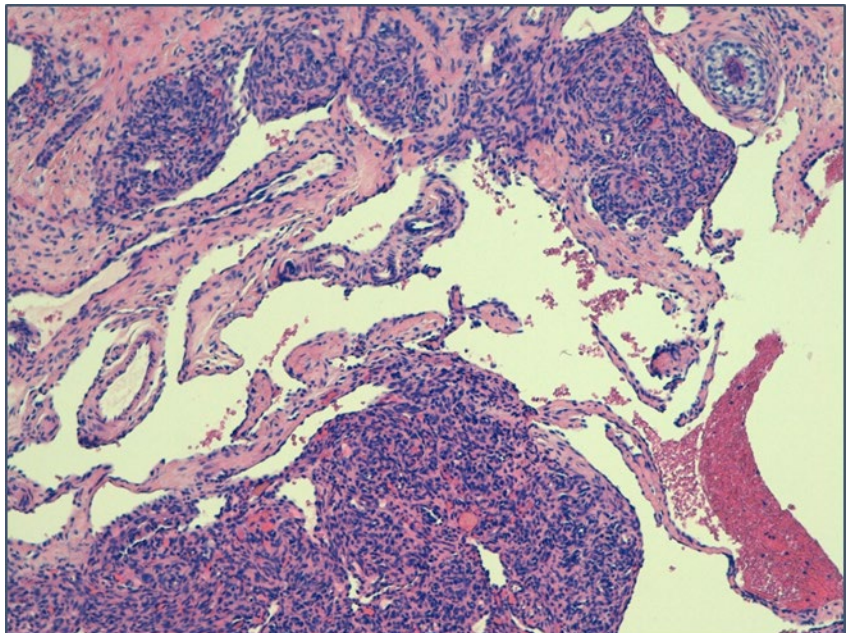


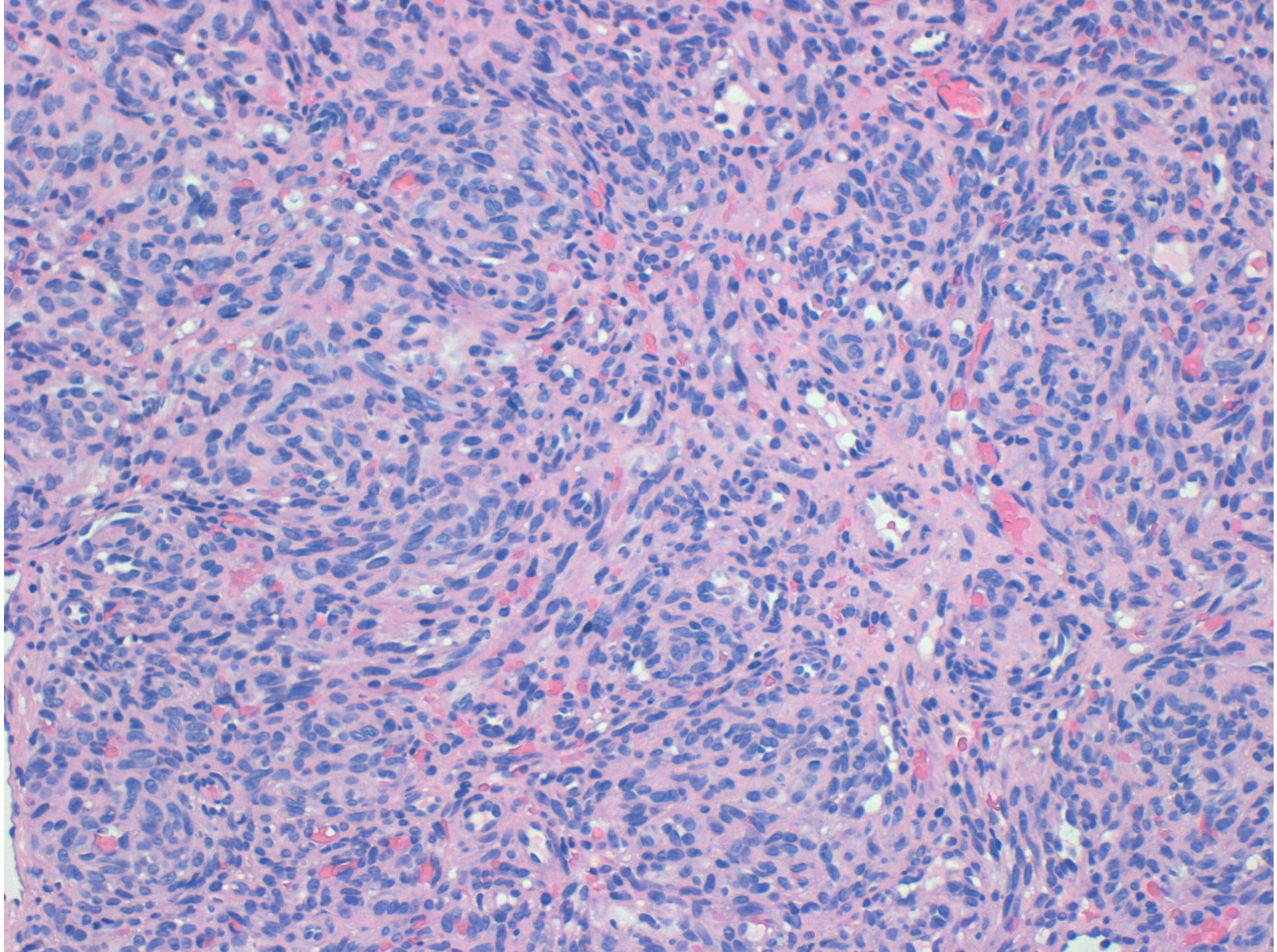


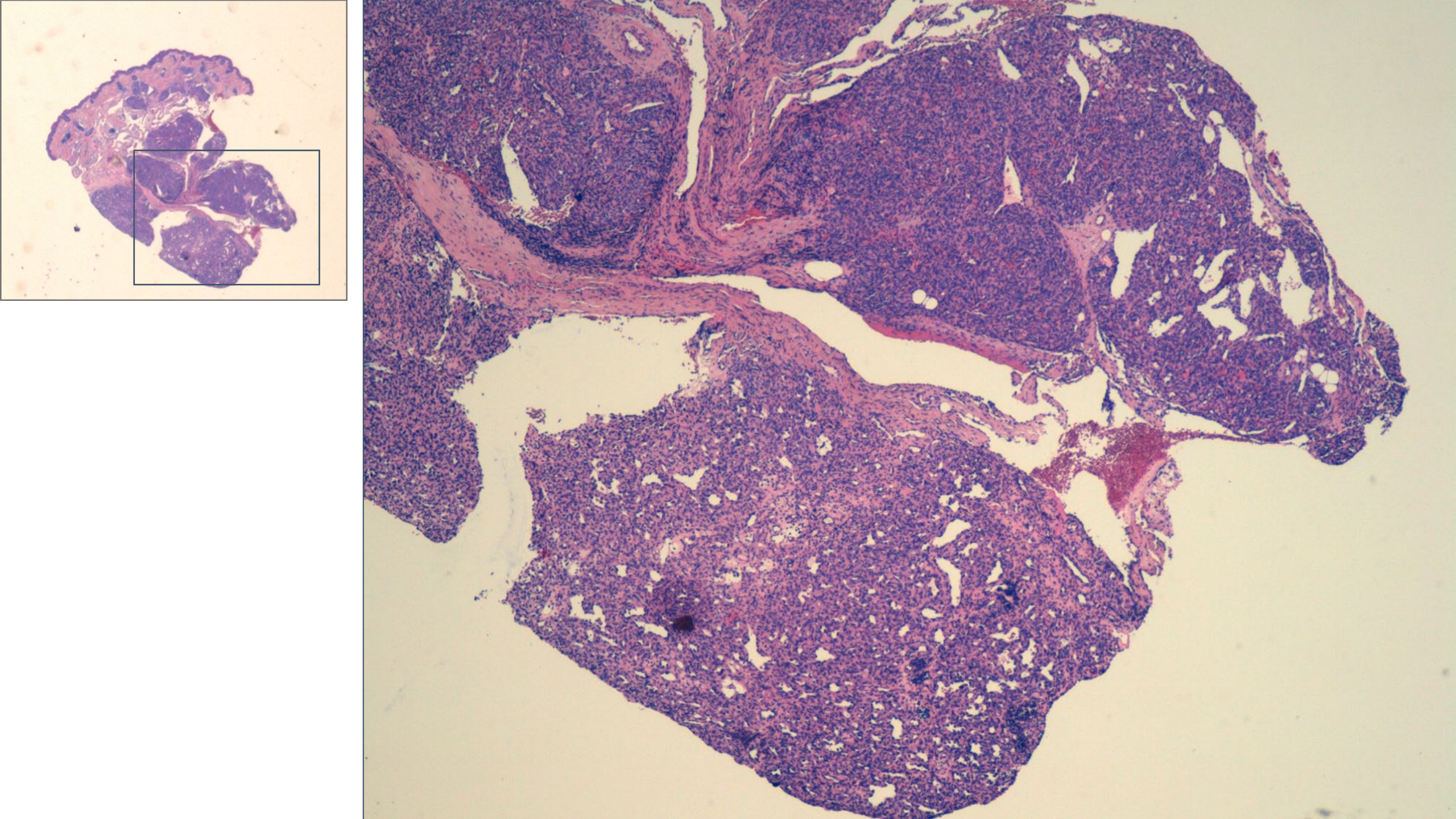


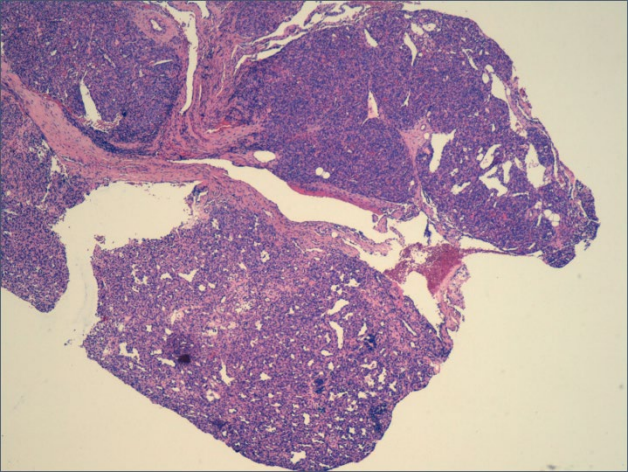


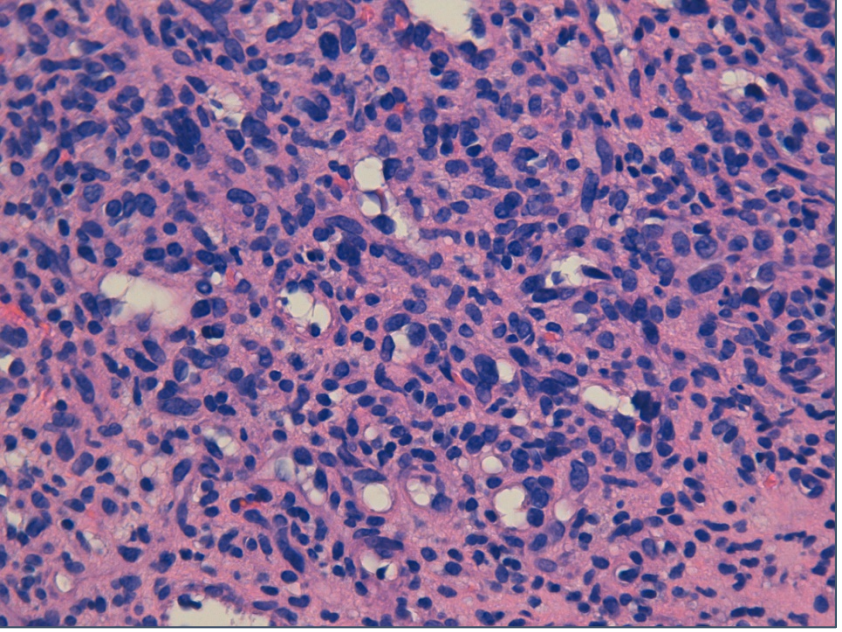
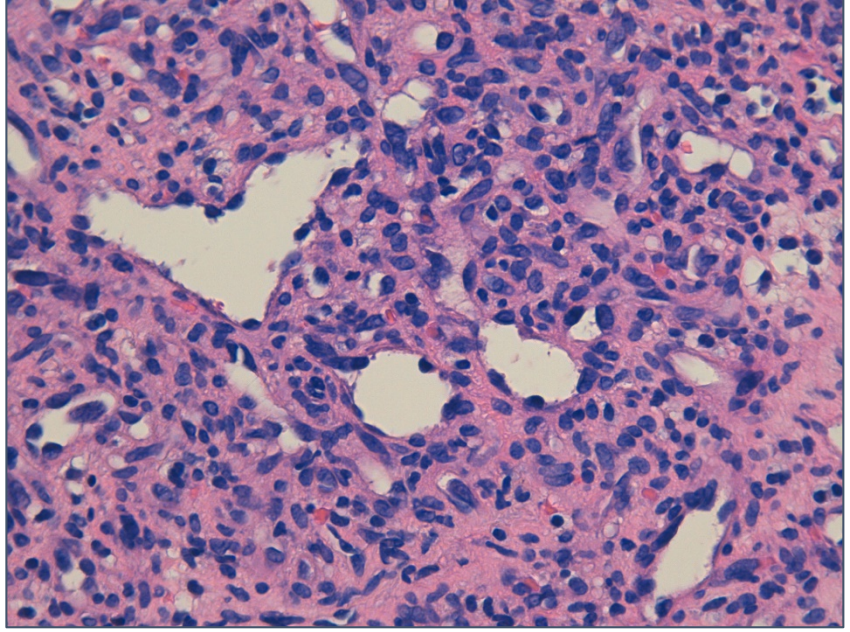
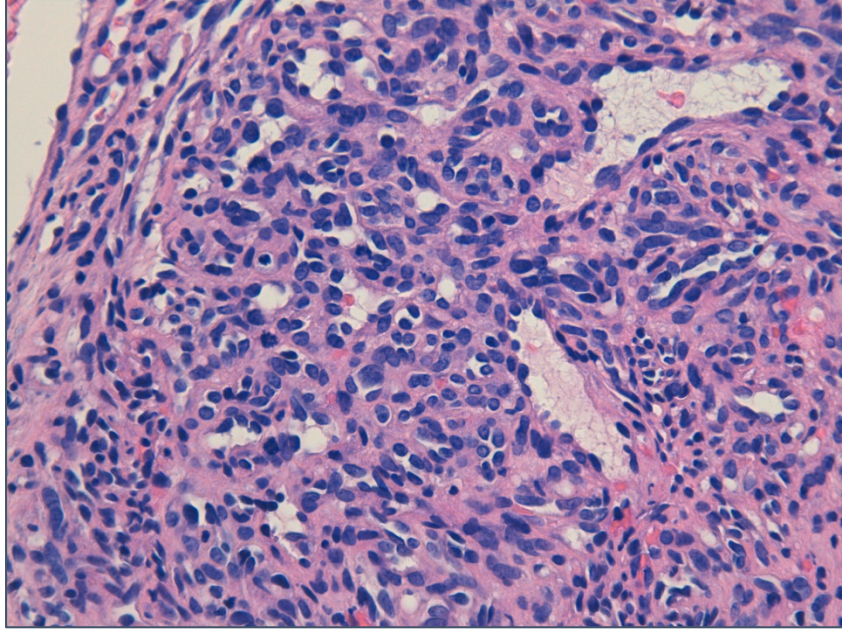
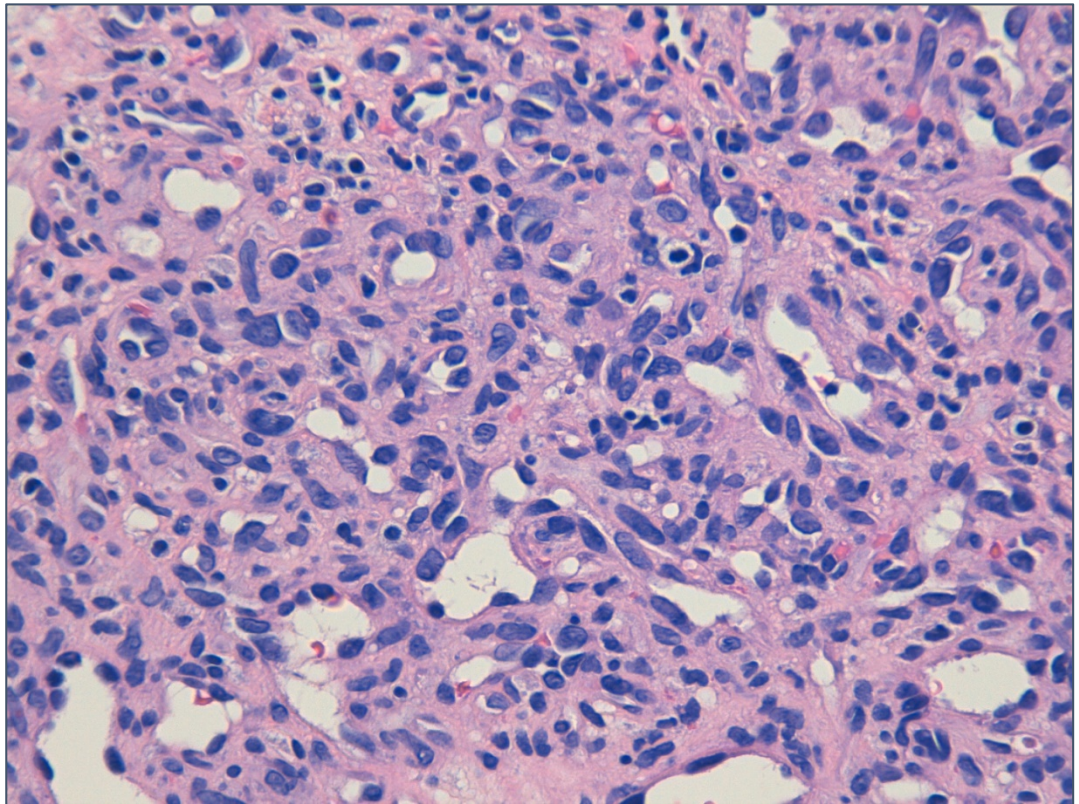
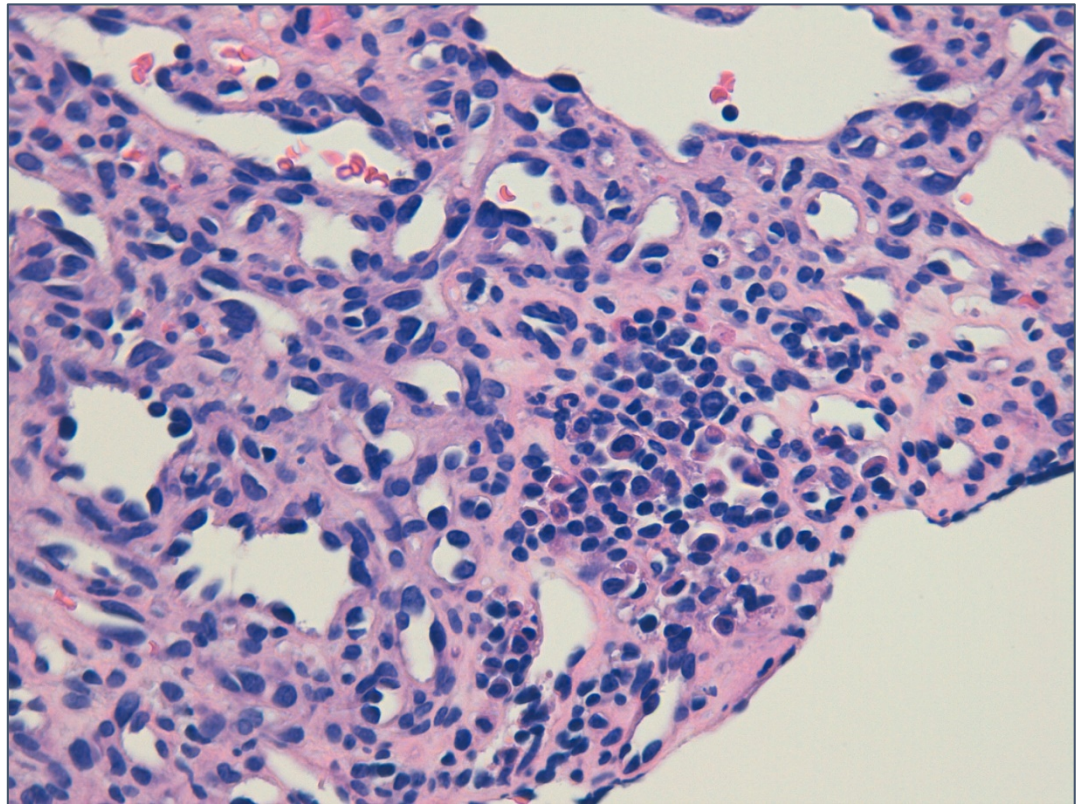


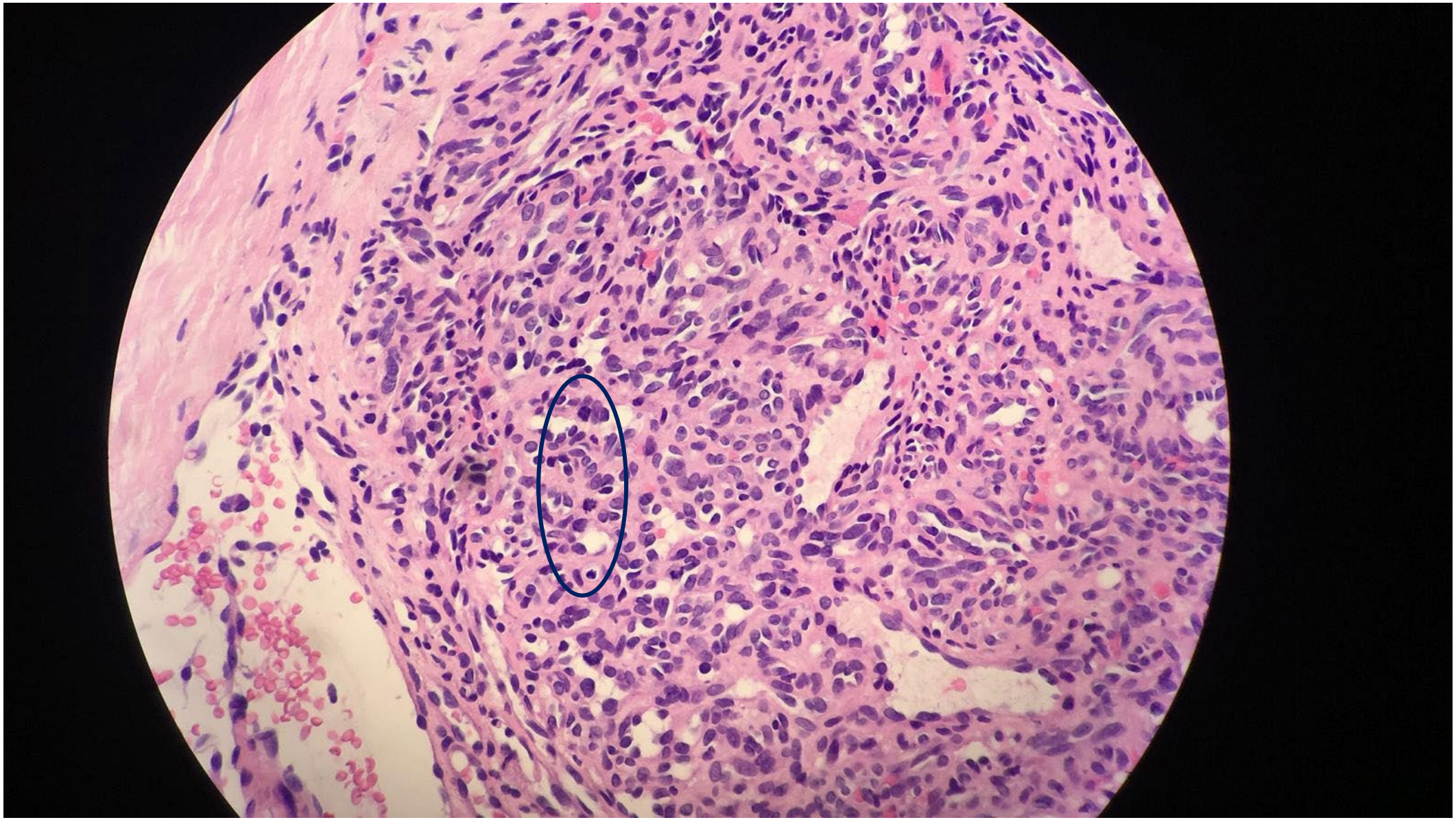


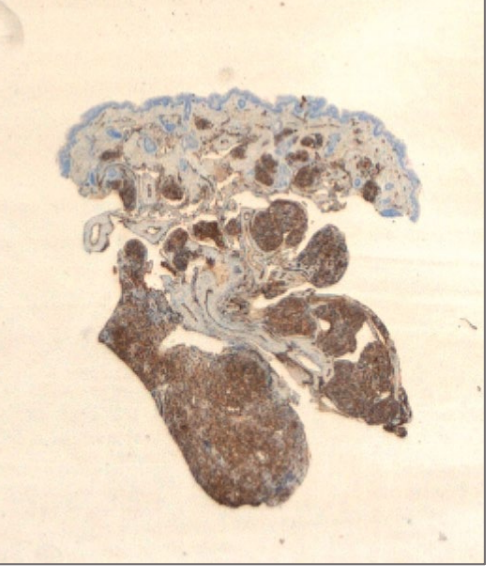


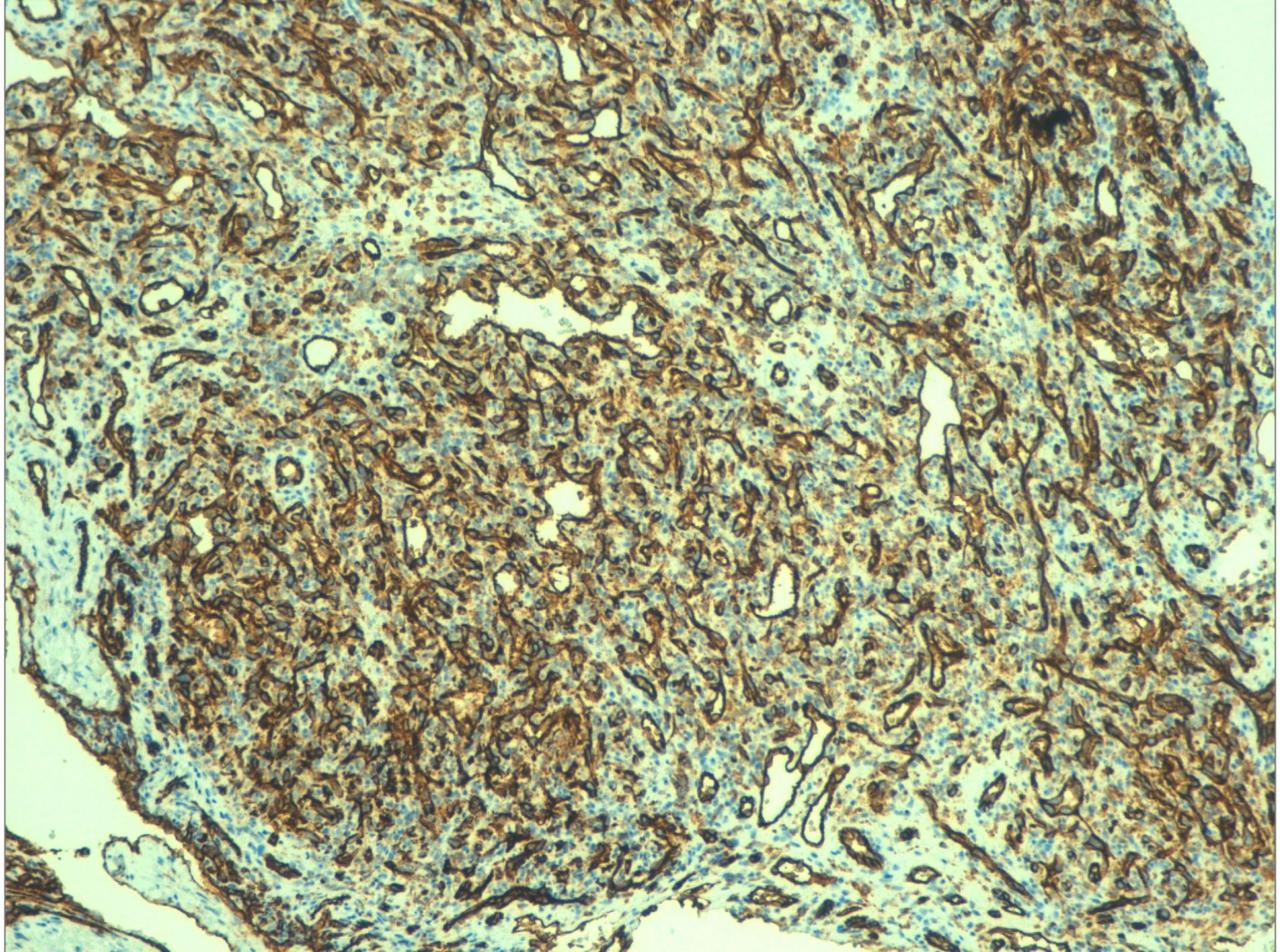






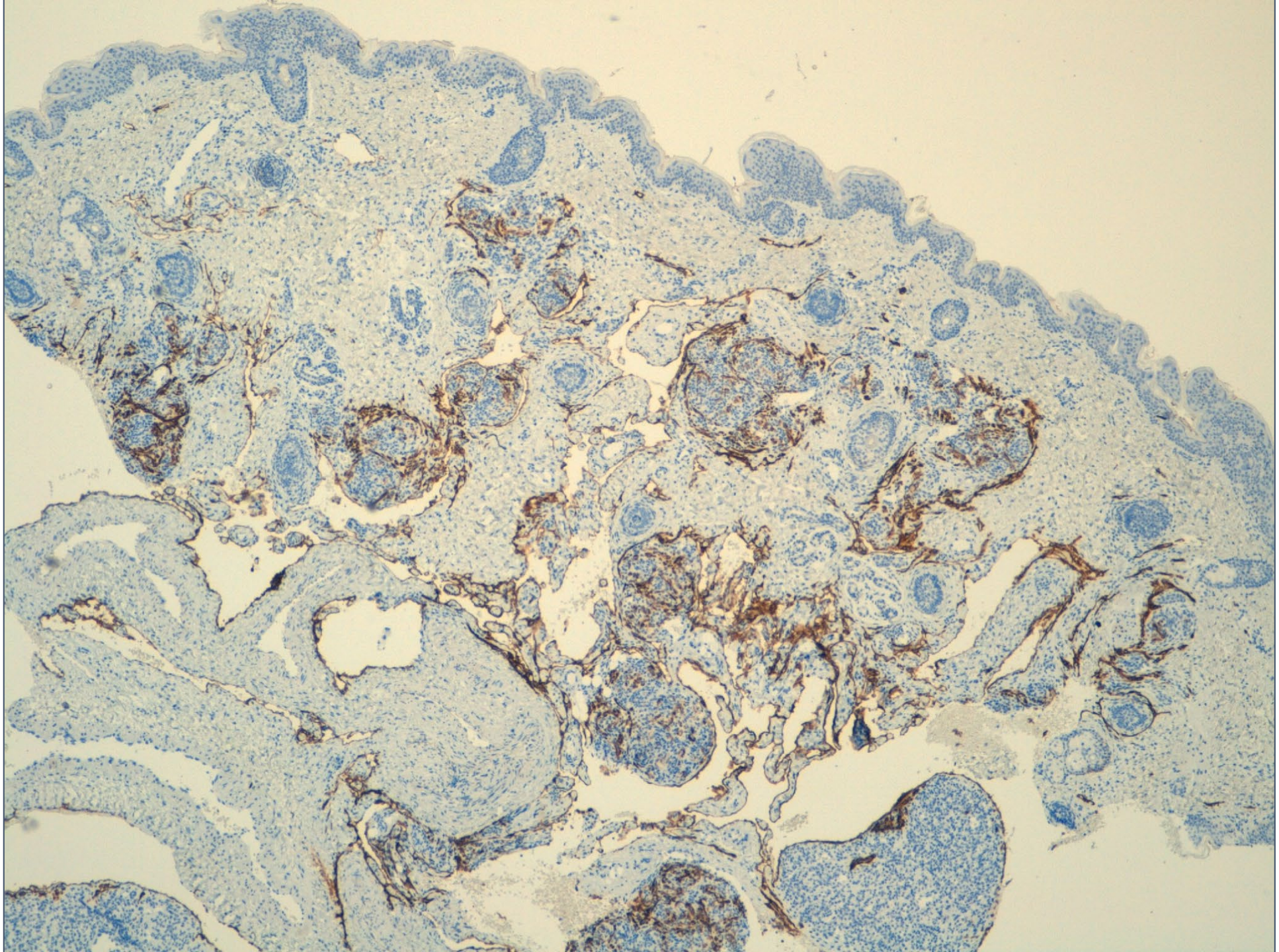


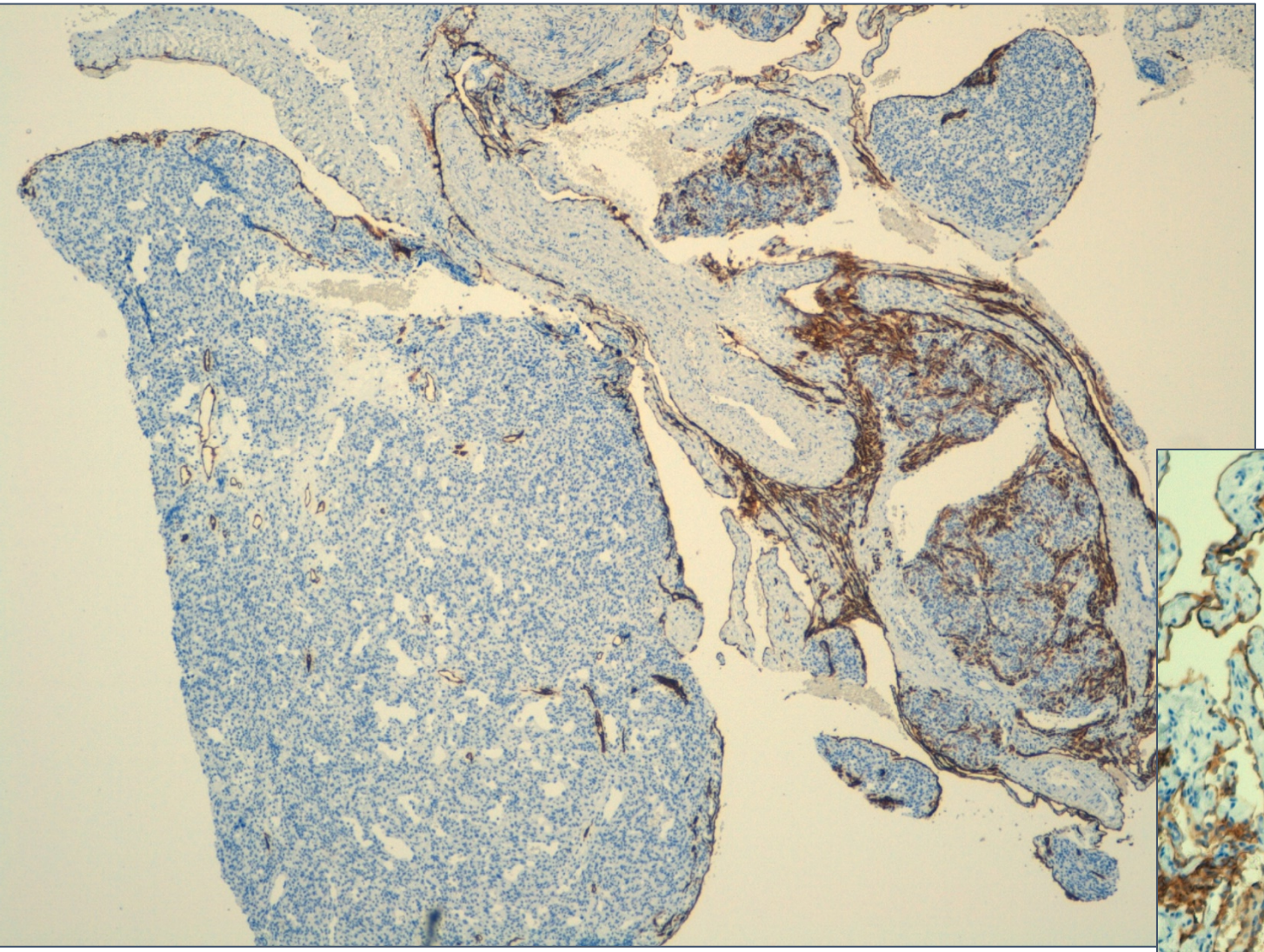


CD31

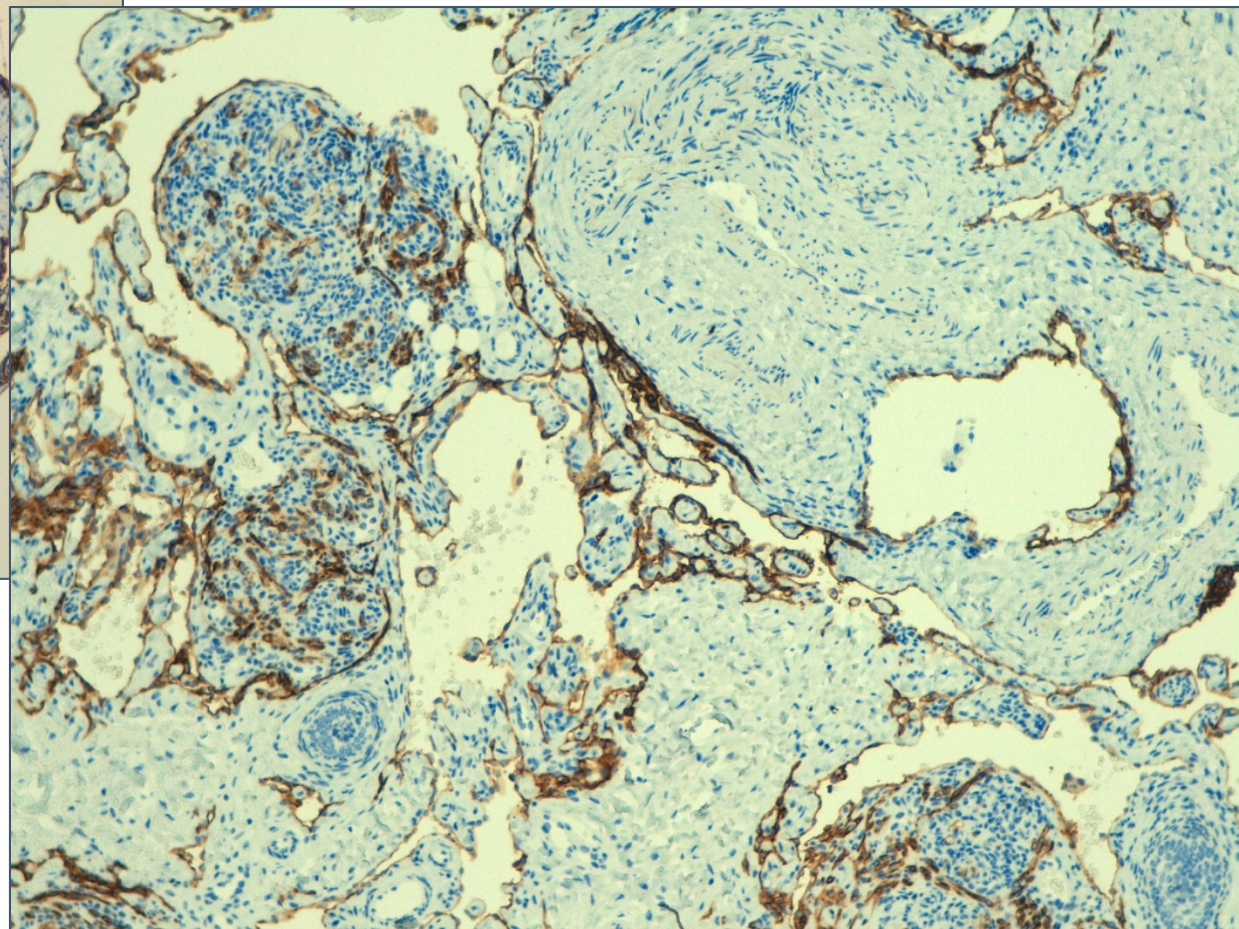
CD34

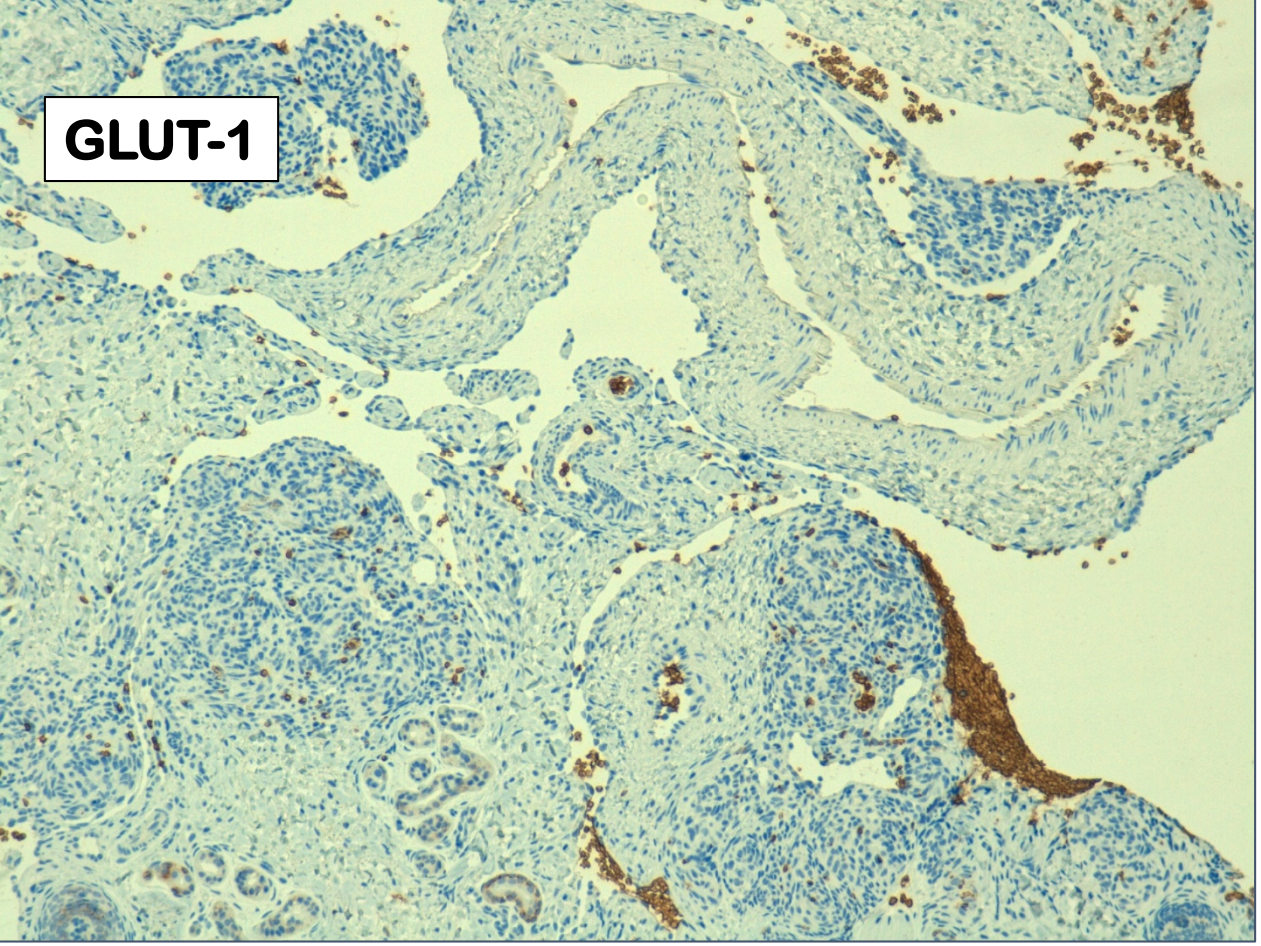
D2-40

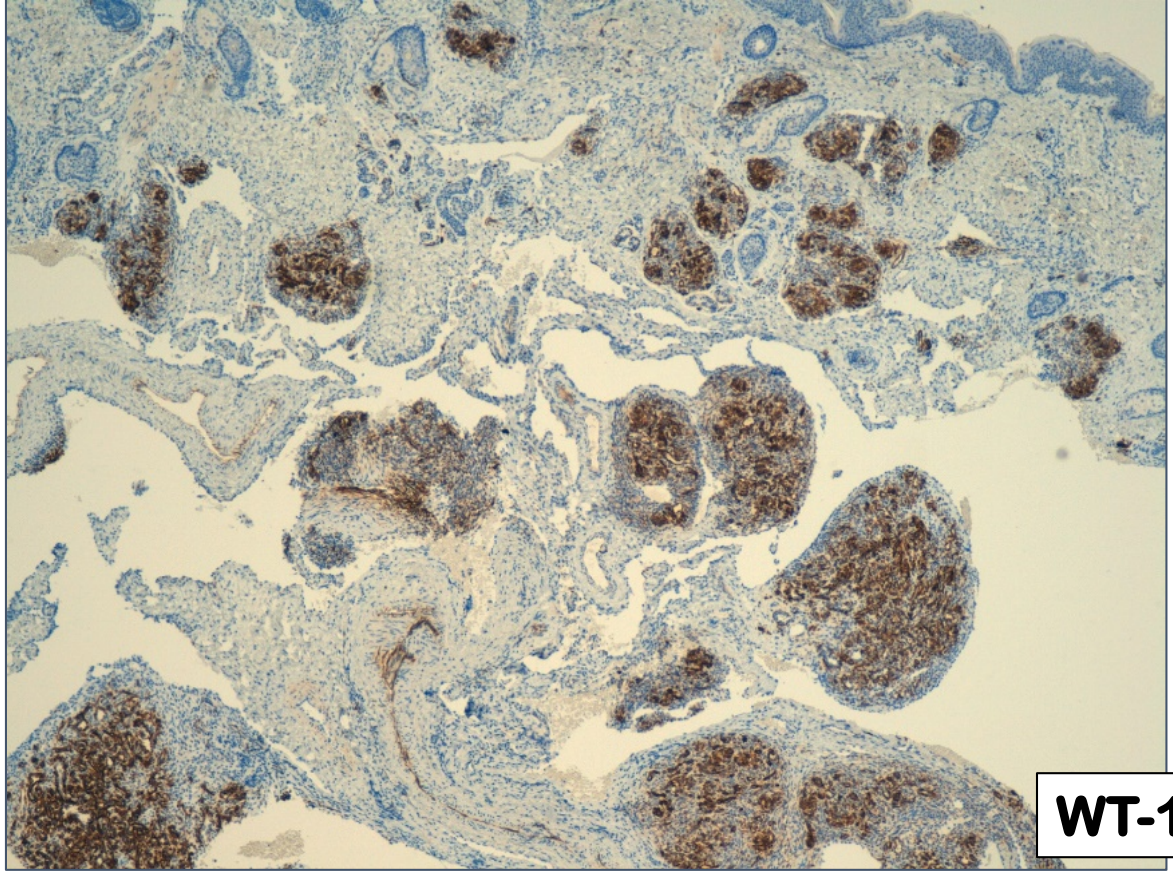


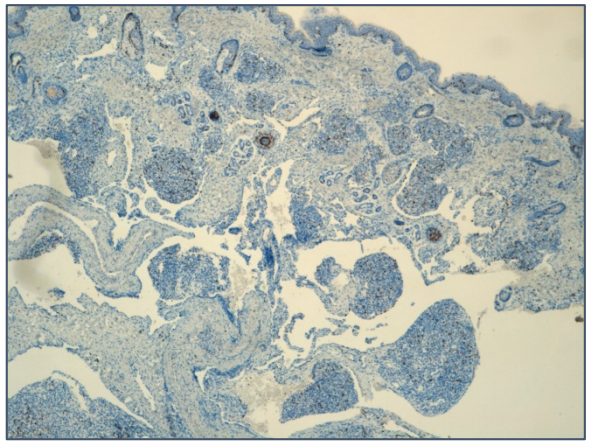


D2-40

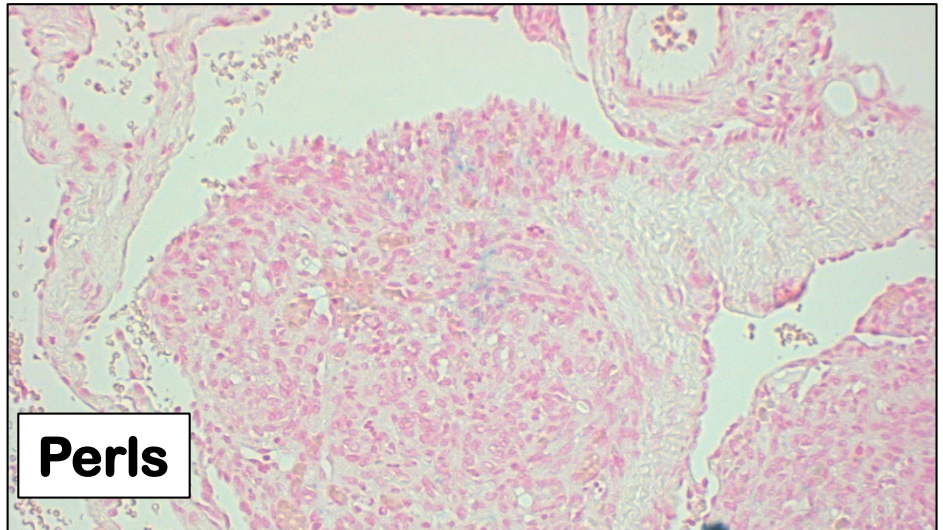








Ki-67



HHV8 -

Tufted Angioma / Kaposiform Hemangioendothelioma

Analítica:

↑ **dímero D, alt. coagulación sin consumo de plaquetas**

RM cervical:

Área de hiperseñal en T2 de bordes mal definidos con afectación difusa de los planos grasos de la región submandibular derecha de 2.4 x 4.3 x 1.2 cm.

Back to overview

ISSVA classification of vascular tumors 1a

Type Alt ← for previous view

Benign vascular tumors 1	
Infantile hemangioma / Hemangioma of infancy	see details
Congenital hemangioma Rapidly involuting (RICH) * Non-involuting (NICH) Partially involuting (PICH)	GNAQ / GNA11
Tufted angioma * °	GNA14
Spindle-cell hemangioma	IDH1 / IDH2
Epithelioid hemangioma	FOS
Pyogenic granuloma (also known as lobular capillary hemangioma)	BRAF / RAS / GNA14
Others	see details

* some lesions may be associated with thrombocytopenia and/or consumptive coagulopathy see details
° many experts believe that tufted angioma and kaposiform hemangioendothelioma are part of a spectrum rather than distinct entities

Back to overview

ISSVA classification of vascular tumors 2

Type Alt ← for previous view

Locally aggressive or borderline vascular tumors	
Kaposiform hemangioendothelioma * °	GNA14
Retiform hemangioendothelioma	
Papillary intralymphatic angioendothelioma (PILA), Dabska tumor	
Composite hemangioendothelioma	
Pseudomyogenic hemangioendothelioma	FOSB
Polymorphous hemangioendothelioma	
Hemangioendothelioma not otherwise specified	
Kaposi sarcoma	
Others	
Malignant vascular tumors	
Angiosarcoma	(Post radiation) MYC
Epithelioid hemangioendothelioma	CAMTA1 / TFE3
Others	

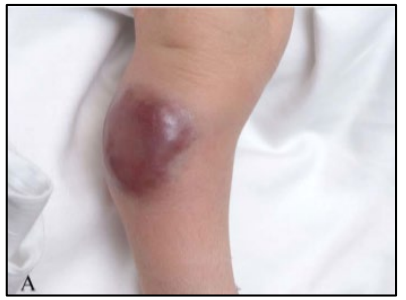
* some lesions may be associated with thrombocytopenia and/or consumptive coagulopathy see details
° many experts believe that tufted angioma and kaposiform hemangioendothelioma are part of a spectrum rather than distinct entities

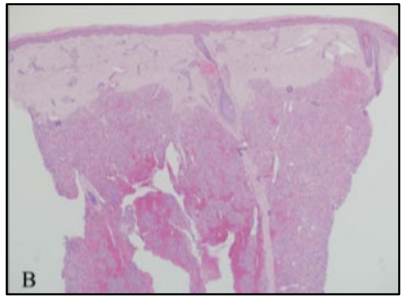
Vascular Tumors in Infants: Case Report and Review of Clinical, Histopathologic, and Immunohistochemical Characteristics of Infantile Hemangioma, Pyogenic Granuloma, Noninvoluting Congenital Hemangioma, Tufted Angioma, and Kaposiform Hemangioendothelioma

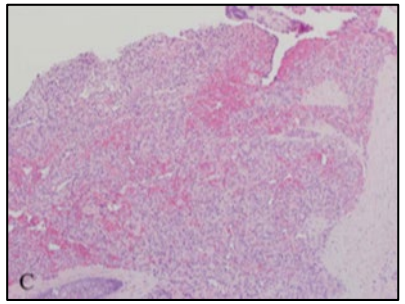
Emma F. Johnson, MD, Dawn M. Davis, MD,†‡§ Megha M. Tollefson, MD,†‡ Karen Fritchie, MD,¶ and Lawrence E. Gibson, MD‖***

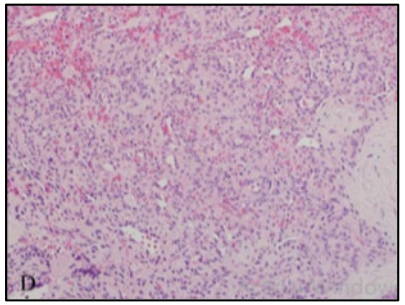
Entity	Histopathology
IH	Dermal proliferation of lobules and sheets of tightly packed, capillary-sized vessels lined with plump endothelial cells. Moderate mitotic figures are present. Mast cells and dermal dendrocyte frequent within stroma.
Pyogenic granuloma	Exophytic, lobulated, dermal mass. Numerous small capillaries with bland endothelial cells in well-developed lobular architecture, variable superficial acute and chronic inflammatory cells.
NICH/RICH	Lobules of congested capillaries surrounded by layer of pericytes separated by bands of fibrosis. Variably atrophic epidermis and adnexal structure. Involving subcutaneous tissue and dermis
TA	Typical "cannonball" distribution of lobules of closely packed poorly canalized capillaries lined with bland endothelial cells and surrounded by pericytes. Involving the dermis and superficial subcutis. Dilated crescent-shaped/semilunar lymphatic channels at periphery.
KHE	Lobular, infiltrative fascicles of endothelial cells, congested capillaries, slit-like vascular spaces, and epithelioid endothelial cells. Capillary may show thrombosis.

Entity	GLUT-1	WT-1	CD31	CD34	D2-40	LYVE-1	Prox-1
IH	+	+	+	+	−	+	−
Pyogenic granuloma	−	+	+	+	−	−	−
NICH	−	+	+	+	−	−	−
TA	−	+	+	+	+ (focally)	+ (focally)	+
KHE	−	NR	+	+ (focally)	+	+ (focally)	+









Histopathology of Spindle Cell Vascular Tumors

Zlatko Marušić, MD, PhD[a], Steven D. Billings, MD[b,*]

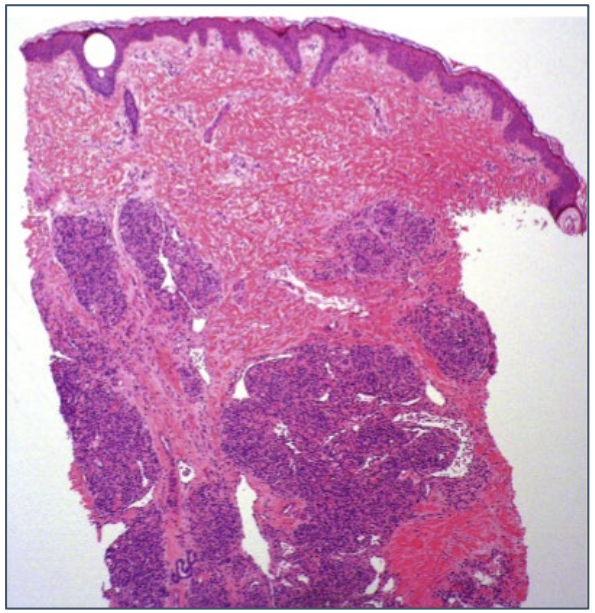
As mentioned previously, acquired tufted angioma and kaposiform hemangioendothelioma represent different spectral parts of the same entity. Both tumors show histologic overlap and a similar immunohistochemical profile. However, lesions reported as acquired tufted angiomas are described as having more of a cannonball pattern and superficial involvement of peripheral soft tissue, whereas kaposiform hemangioendothelioma is deeper and more extensive. Acquired tufted angiomas also show lower propensity for the induction of Kasabach-Merritt syndrome, but this may simply be a reflection of small tumor size in entities diagnosed as acquired tufted angioma.[2]

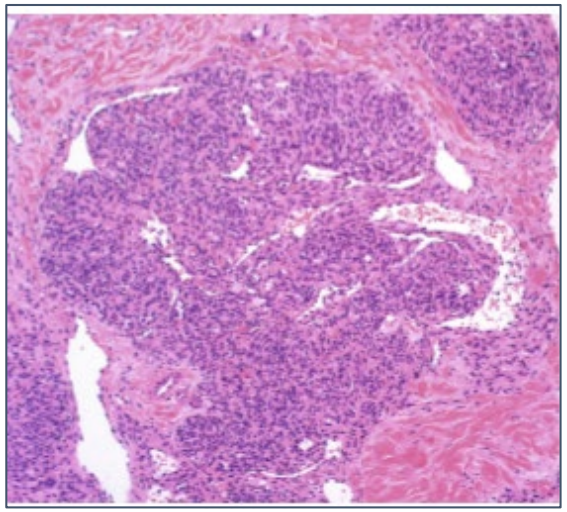
Pitfalls
Tufted Angioma

! Shows considerable overlap with kaposiform hemangioendothelioma

! The term tufted angioma should probably be reserved for small cutaneous lesions in adults

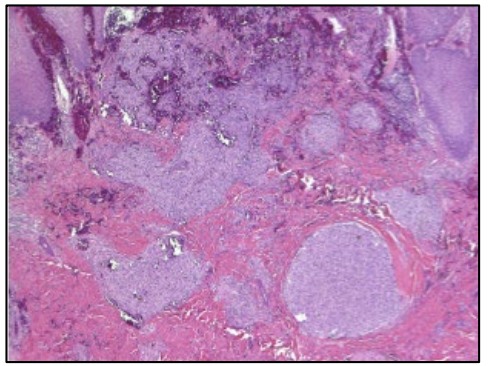
gioendothelioma. However, they likely represent a spectrum of the same tumor. It may be reasonable to retain the terminology of acquired tufted angioma for small, superficial lesions, especially those presenting in adult patients, reserving the diagnosis of kaposiform hemangioendothelioma for the more extensive and deeper childhood lesions associated with consumptive coagulopathy.

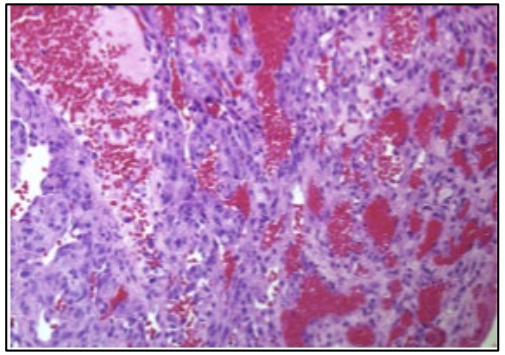


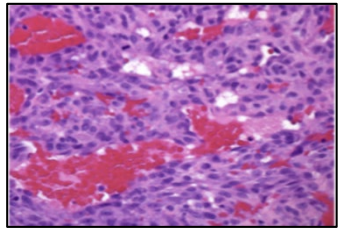


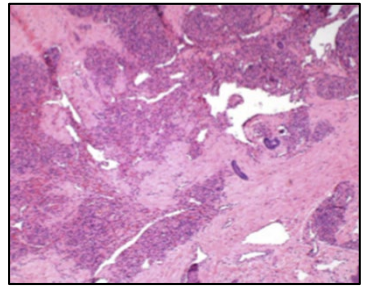
Acquired Tufted Angioma vs	Helpful Distinguishing Features
Kaposiform hemangioendothelioma	• Likely a spectrum of the same tumor • More extensive and confluent growth • More frequent Kasabach-Merritt syndrome
Nodular Kaposi sarcoma	• Lacks the cannonball pattern • More confluent spindle cell population and diffuse reticulin network pattern • Invariably positive for HHV-8

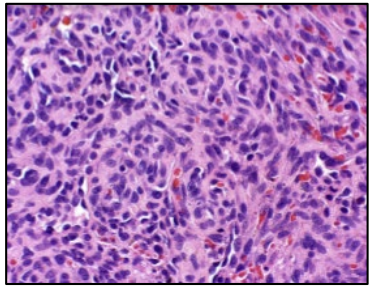
vs







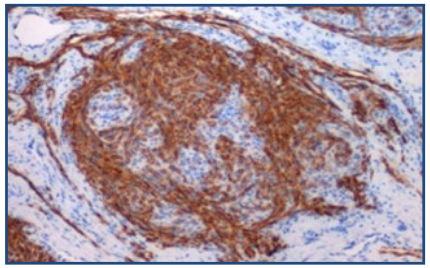


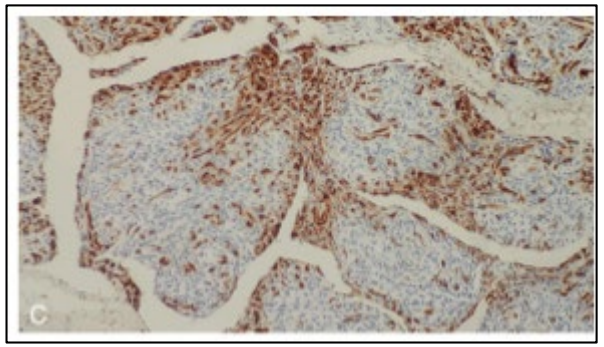


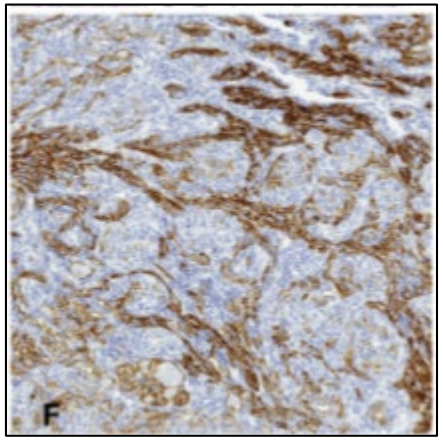
Mitosis:

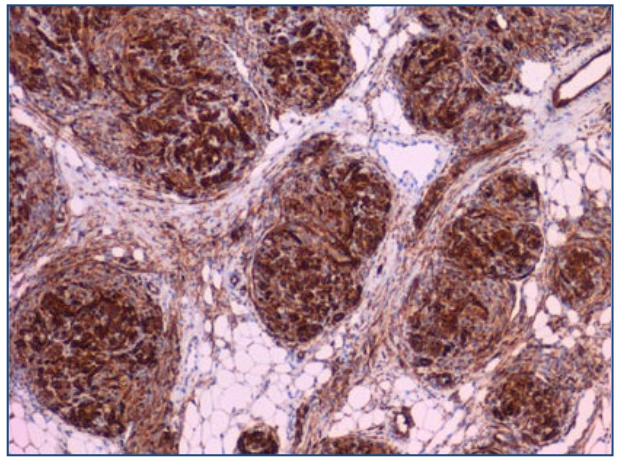
Histologically, the tumors were dominated by infiltrating isolated and fusional nodules of spindle cells forming characteristic vascular pattern. The tumors of 11 cases were presented with isolated nodules mainly, 6 cases with fusional nodules, 5 cases with mixed pattern. The tumor nodules were composed of fascicles of spindleshaped endothelial cells and slitlike or crescentic vascular channels. Most cases (14 cases, 64%) had unclear and irregular tumor margins. In addition, there were extravasated red blood cells (RBC's), single cells with lumina containing RBC's, fibrin thrombi, eosinophilic globules and hemosiderin. There was mild nuclear variation, but no significant nuclear atypia, or necrosis, with rare mitoses (0-8/10HPF, mean 2/10HPF). In 2 cases, the tumors involved the skin nervelet. In all cases the stroma was predominantly collagenous, and discrete foci of irregular dilated vascular and lymphatic vessels. Only 1 case had a background of extensive lymphangiomatosis-like changes. And in another case there were focus calcification and lymphocytic infiltration in the stroma. It was be found that the trapping of RBC's and lymphocytes in the slitlike channels of the tumor nodules (Figure 3).

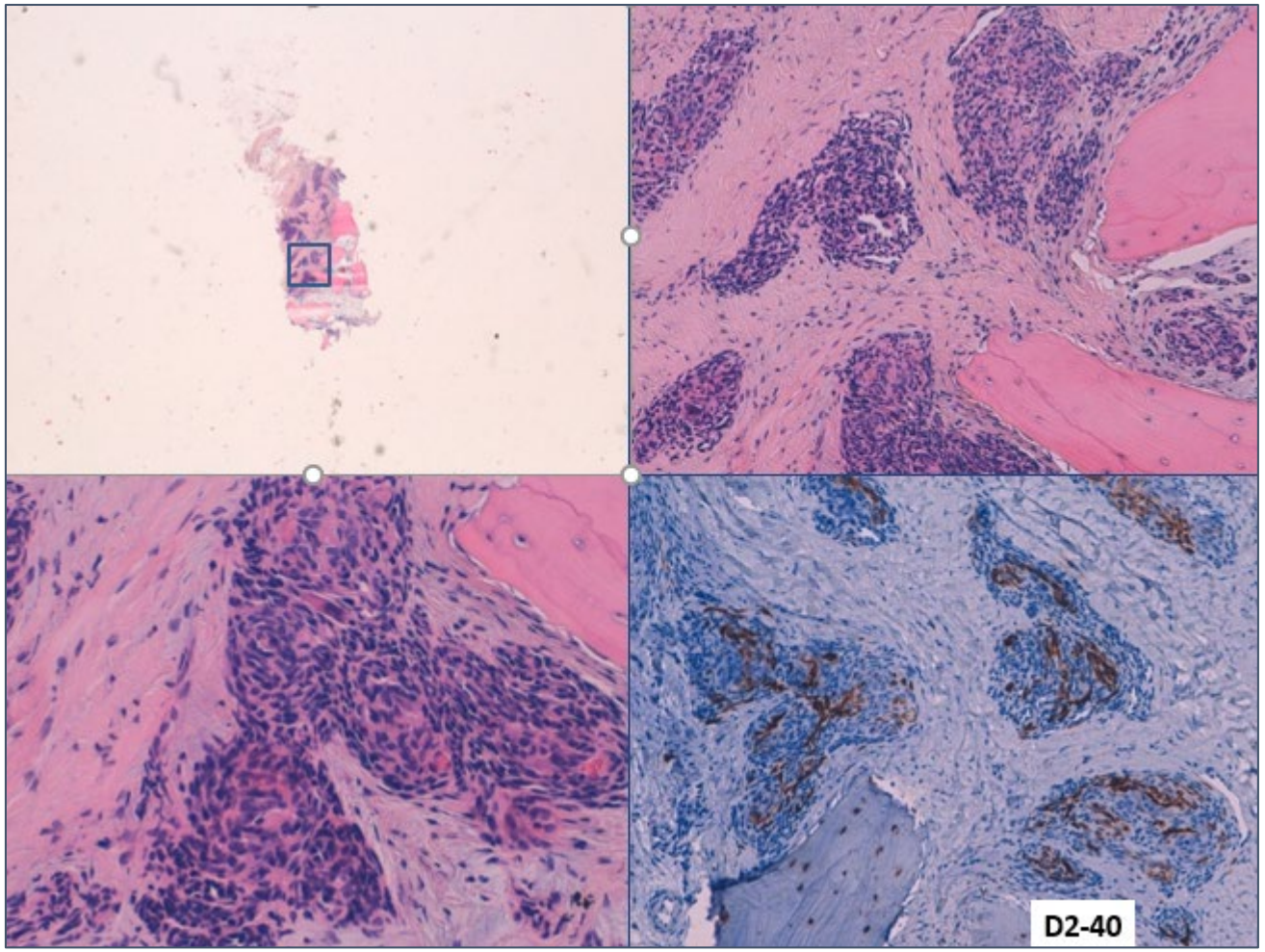
Expresión de D2-40:











Sirolimus in the Treatment of Vascular Anomalies

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Eur J Pediatr Surg 2017;27:86–90.

Eur J Pediatr Surg. 2017 Feb;27(1):86-90. doi: 10.1055/s-0036-1593383. Epub 2016 Oct 10.

Sirolimus in the Treatment of Vascular Anomalies.

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Author information

Abstract

Aim of the Study mTOR inhibitors are showing promising results in the management of vascular anomalies. Although current controlled trials remain to be completed, many individual experiences are being published. We present our series of children with complex vascular anomalies treated with sirolimus. **Patients and Methods** A retrospective review of 41 patients treated with sirolimus between January 2011 and December 2015 was performed: 15% ($n = 6$) had vascular tumors (4 kaposiform hemangioendotheliomas, 1 PTEN) and 85% ($n = 35$) had malformations (13 generalized lymphatic anomalies/Gorham-Stout diseases [GSD], 1 kaposiform lymphangiomatosis [KLA], 11 large lymphatic malformations (LMs) in critical areas, 2 lymphedemas, 4 venous malformations, and 4 aggressive arteriovenous malformations [AVM]). Several variables were collected: type of vascular anomaly, duration of treatment, dosage, response, and secondary effects. **Results** There was a female predominance (1.4:1). All patients received sirolimus, at initial dosage of 0.8 mg/m^2/12 hour. Overall successful response rate was 80.4% of cases, presenting improvement in radiologic imaging and reduction of symptoms, at a median time of 10 weeks. Patients showing no response included four AVMs, one GSD, one LM, one KLA, and one unknown tumor. Sirolimus was well tolerated, even in neonates, with insignificant side effects. No patients had complete resolution and no patients worsened on therapy. Thirty patients remain under treatment at the present moment. **Conclusion** Sirolimus has become a new therapeutic option for patients with vascular anomalies that do not respond to other treatments. Unfortunately, important questions as what is the most appropriate dosage and for how long should the patient be treated remain unanswered. An international registry followed by customized controlled trials is mandatory to clarify the future of this therapy.

CASE REPORT

Sirolimus for treatment of kaposiform hemangioendothelioma associated with Kasabach-Merritt phenomenon

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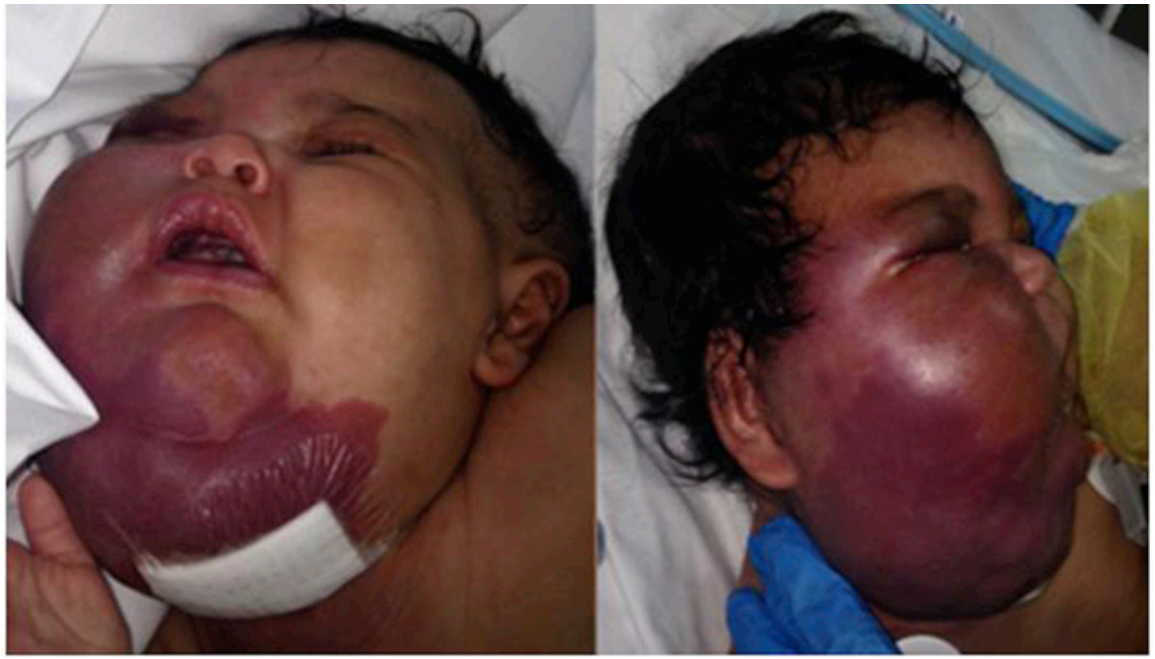
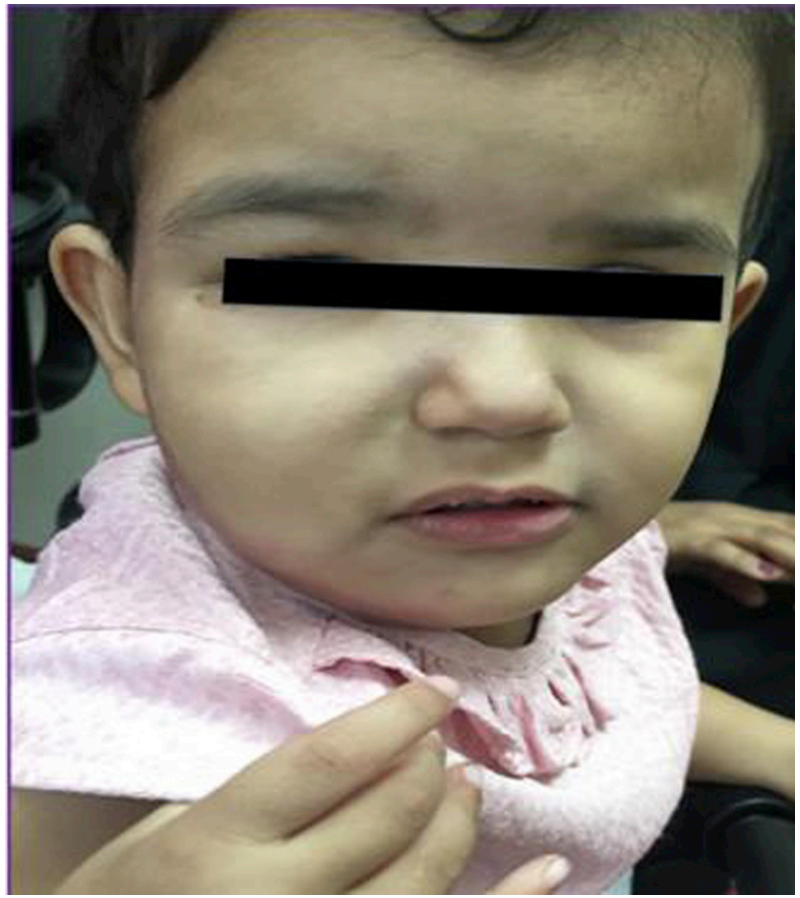
Key words: case report; kaposiform hemangioendothelioma; Kasabach-Merritt phenomenon; sirolimus; treatment; vascular.





Pancreatic Kaposiform Hemangioendothelioma Not Responding to Sirolimus

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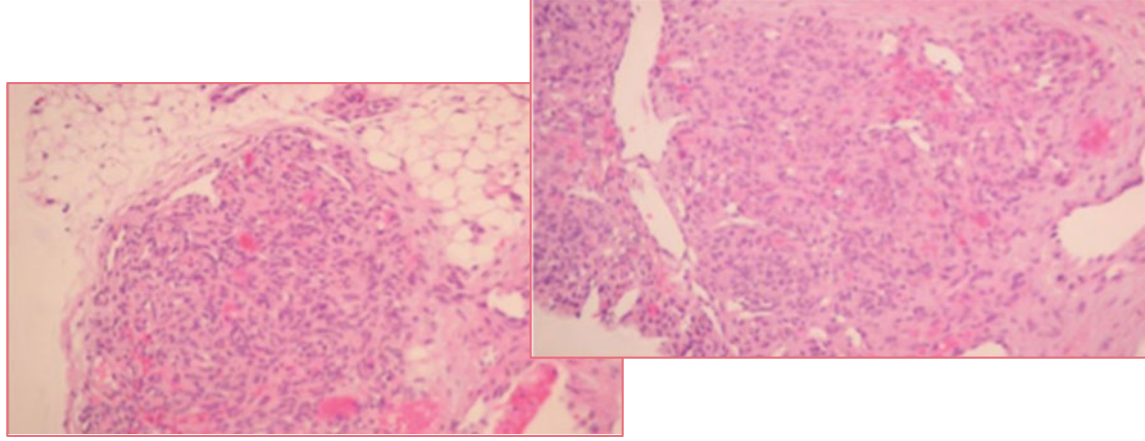
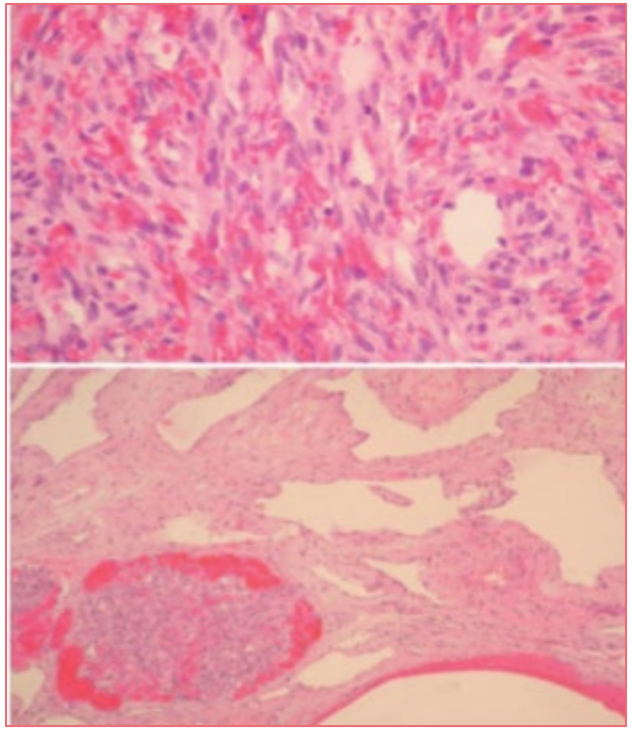
Eur J Pediatr Surg Rep 2017;5:e32–e35.





Abstract

Background Kaposiform hemangioendothelioma (KHE) is a vascular tumor frequently associated with Kasabach–Merritt phenomenon (KMP), characterized by severe thrombocytopenia and consumptive coagulopathy. Visceral involvement in KHE is rare. In our recent experience, sirolimus has shown to be an effective treatment in cutaneous KHE, becoming indeed the treatment of choice in KMP. We report a case of pancreatic KHE associated with KMP and refractory to sirolimus.

Case Report A 4-month-old infant is referred for obstructive jaundice (10 mg/dL conjugated bilirubin) secondary to vascular pancreatic tumor. Magnetic resonance (MR) and immunohistochemistry were compatible with KHE, but the tumor was considered unresectable. We initiated sirolimus (0.8 mg/m^2/12 h) to treat KMP, and interventional radiology was performed for percutaneous biliary diversion. This procedure prompted KMP (platelets: 51,000/µL). Sirolimus treatment for 7 days showed no effect; therefore, we started our VAT protocol (vincristine/aspirine/ticlopidin) with great response after 10 days (platelets: 3,70,000/µL). Three months later, percutaneous biliary diversion was replaced by a biliary stent. The tumor disappeared leaving fibrosis and dilatation of biliary tract needing hepaticojejunostomy 6 months later.

Discussion It is difficult to establish protocols for an unusual presentation of a tumor with different targets. This is a reason collaborative multicenter studies should be performed. Management of obstructive jaundice secondary to a tumor that usually regresses in 10 years is an added challenge; therefore, the management should be led by a multidisciplinary team.

Sirolimus treatment in cutaneous KHE has been described as successful in the literature, as well as in our own experience; however, it failed in our first patient with visceral KHE. We need to investigate the different response to pharmacological agents in tumors with similar histopathology, but with visceral involvement.

Keywords

- kaposiform hemangioendothelioma
- pancreatic
- Sirolimus

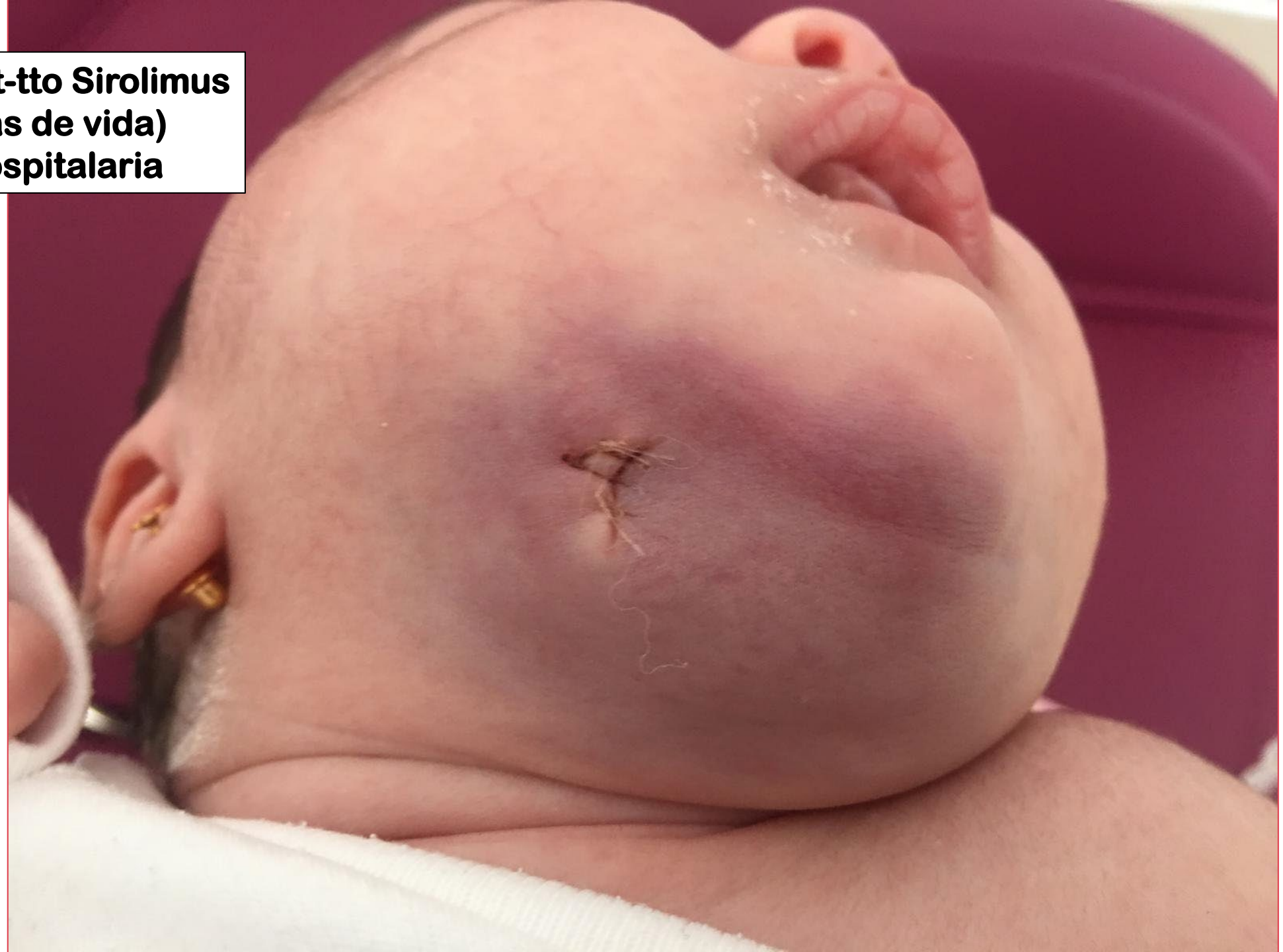
IC

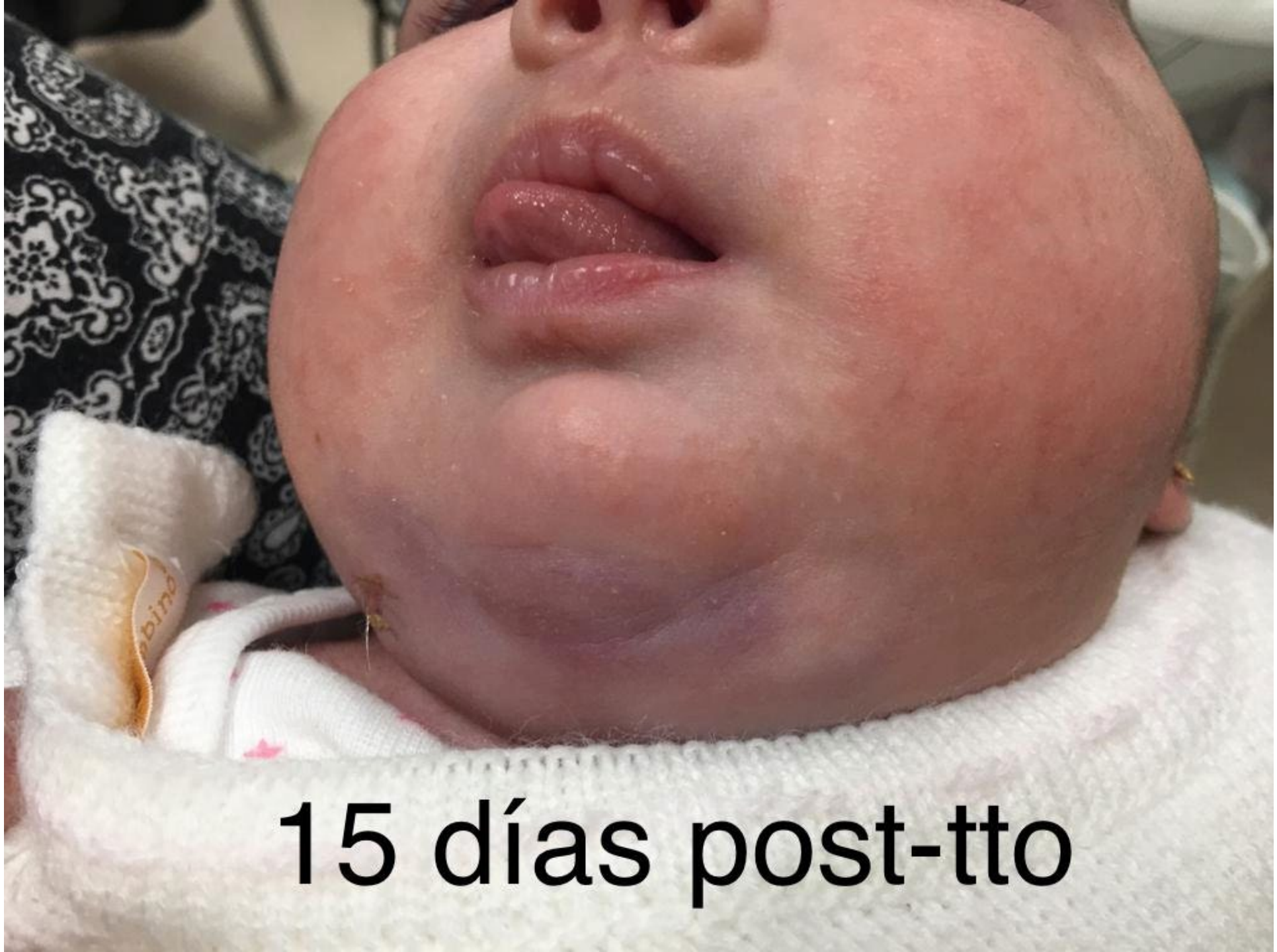
Isabel Colmenero <isabelcolmenero@gmail.com>
Jue 29/11/2018, 3:51
Usted

Totalmente de acuerdo, tiene de ambos, lo cual no es muy infrecuente. Es difícil saber cuánto tiene de KH porque es una biopsia superficial, pero los nódulos grandes fusionados definitivamente son de KH. La superficie es más de TA.
En realidad lo más importante aquí es tratarle el KM y eso ya está hecho.
Yo lo daria como tú dices, de tumor de la familia (o espectro) TA/KH.
Un abrazo,
IC

Sent from my iPhone

4º día post-tto Sirolimus
(41 días de vida)
Alta hospitalaria




15 días post-tto


GRACIAS