

CASE REPORT

Lipomatous hemangiopericytoma in a child: A case report with immunohistochemical evaluation

Jayakiran Madala, Venkateswara Rao Guttikonda, Rajani Korlepara

Department of Oral Pathology and Microbiology, Mamata Dental College, Khammam, Andhra Pradesh, India

Address for correspondence:

Dr. Jayakiran Madala,
Department of Oral Pathology and Microbiology,
Mamata Dental College, Giriprasad Nagar,
Khammam - 507 002, Andhra Pradesh, India.
E-mail: jaikiran78@gmail.com

Received: 25-10-2013

Accepted: 23-03-2015

ABSTRACT

Lipomatous hemangiopericytomas are rare soft-tissue tumors that show areas of hemangiopericytoma like vascular pattern admixed with lipid containing cells. It is now considered a subtype of solitary fibrous tumor due to histopathological and immunohistochemical similarities. To date, only a few cases have been adequately reported in the medical literature. A case of lipomatous hemangiopericytoma in an 11-year-old female patient is presented and the clinical, histopathological and immunohistochemical features are discussed, along with a literature review. To the best of our knowledge, this is the first known case of a lipomatous hemangiopericytoma in a pediatric patient. Our case report further emphasizes that a number of studies should be evaluated to delineate hemangiopericytomas from solitary fibrous tumors.

Key words: Hemangiopericytoma, lipomatous hemangiopericytoma, solitary fibrous tumor

INTRODUCTION

Hemangiopericytoma (HPC) is a rare tumor, first described in 1942 by Stout and Murray comprising of capillary blood vessels and proliferating perivascular round cells, warranting hemangiopericytoma as a proper descriptive name.^[1] It has been defined as a tumor composed of spindle to oval shaped undifferentiated cells, which proliferate around and are intimately associated with prominent, thin walled, often branching vessels.

Hemangiopericytoma is most commonly seen in the fifth-sixth decades of life with only 5–10% of cases occurring in childhood.^[2] The histogenesis and histological classification of HPC is still a controversy, HPC is no longer considered a specific entity, but instead a growth pattern, shared by a variety of unrelated soft tissue neoplasms and should be considered a diagnosis of exclusion.^[3] In the 2002 edition of the WHO classification for soft tissue tumors, HPCs were sub-classified into the group of fibroblastic and myofibroblastic soft tissue tumors. Most of the tumors formerly called HPCs show a more fibroblastic than pericytic differentiation and are now classified as solitary fibrous tumors.^[4]

Solitary fibrous tumor (SFT) is a distinctive neoplasm occurring predominantly in the pleura and was first reported by Kemperer and Rabin *et al.*, in 1931.^[5] Fat-forming variant of solitary fibrous tumor, previously called lipomatous hemangiopericytoma (LHPC), is a recently recognized rare variant of solitary fibrous tumor with unpredictable biologic behavior. Most commonly it occurs in the deep soft tissues of the thigh and retroperitoneum.^[6] Histologically, it is characterized by a prominent hemangiopericytomatous vasculature along with mature adipocytes in the tumor.^[3,6] It usually occurs in middle aged adults with no sex prediction. Only a few cases have been reported in the literature. We present the clinical, radiological and histological features of a case of LHPC in the submandibular region of a pediatric patient.

CASE REPORT

A 11-year-old female presented with a painless large diffuse swelling on the lower right side of the face involving the inferior border of the mandible that had been present for past 2 years. The swelling extended superoinferiorly from 1cm below the ala-tragus line to 1 cm below the lower border of the mandible and anteroposteriorly 2 cm short of the angle of the mandible to angle of the mouth. The margins of the swelling were ill-defined. The tumor was hard and elastic and had a smooth surface [Figure 1]. Intraoral examination revealed obliteration of the lower buccal vestibule in relation to permanent mandibular right canine, extending up to retromolar pad area with no other significant findings.

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10.4103/0973-029X.157209

MRI revealed approximately $50 \times 28 \times 25$ mm altered signal lesion seen on the right side of the mandible with involvement of alveolar surface. The lesion was heterogeneously hyperintense on T2 and hypointense on T1 with small fatty component and cystic focus [Figure 2]. There was no fat plane with adjacent tongue suggesting its involvement. The right sub mandibular gland was abutted. No significant flow voids were noted. Computed tomography (CT) scan showed well defined lobulated heterogenous mass in the right submandibular region extending into ipsilateral sublingual space. Erosion of cortex was also noted.

Wide surgical excision of the tumor was done ensuring safe margins and the patient has been free of any recurrences or metastasis for over a year. Grossly, the lesion removed measured $6 \times 4 \times 4.5$ cm. The lesion was well-circumscribed with yellowish tan in appearance with soft lipid like regions. The lesion was soft and rubbery in consistency. Microscopically, the tumor was well circumscribed and exhibited a lobular growth pattern with prominent HPC like areas admixed with mature adipocytes [Figure 3]. However, no lipoblasts were seen. The HPC regions showed characteristic branched stag-horn vessels, which are lined by flattened endothelial cells. The intervascular

spaces were occupied by monomorphous plump spindle cells which had round to oval nuclei with scant to moderate cytoplasm, the cell borders were indistinct [Figure 4]. There was no significant pleomorphism or mitotic activity. Variable amount of collagen deposition was seen.

Immunohistochemical staining demonstrated that the neoplastic cells were strongly positive for reticulin [Figure 5] and negative for CD 99 [Figure 6], Bcl-2 [Figure 7], S-100 [Figure 8], Pancytokeratin [Figure 9], Desmin [Figure 10], CD34 [Figure 11] and smooth muscle actin (SMA) [Figure 12]. However, CD34 showed positivity in endothelial cells whereas SMA showed positivity in vessel walls. The fat-containing cells exhibited positive staining for S-100.

DISCUSSION

Lipomatous hemangiopericytoma is a rare variant of hemangiopericytoma, composed of mature adipocytes and hemangiopericytomas like areas.^[3,6] In 1995, Nielson *et al.*,



Figure 1: Extraoral photograph showing facial asymmetry with a smooth surfaced swelling on the right lower side of the face

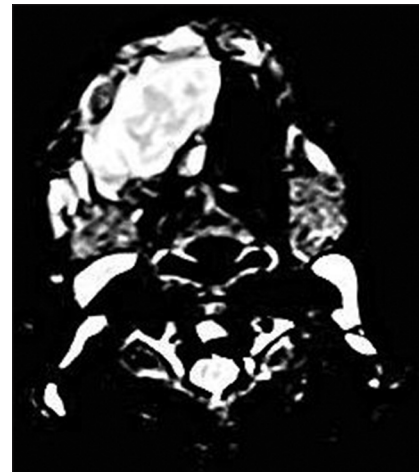


Figure 2: MRI - STIR image showing hypointense areas suggestive of fat

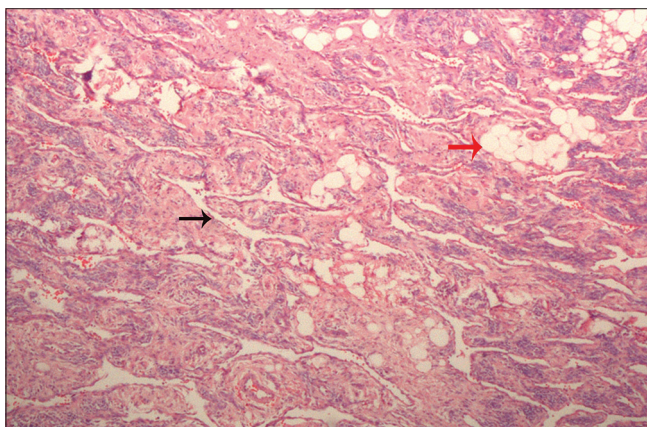


Figure 3: Hemangiopericytoma-like areas (black arrow) admixed with areas of mature adipocytes (red arrow) (H&E stain, x40)

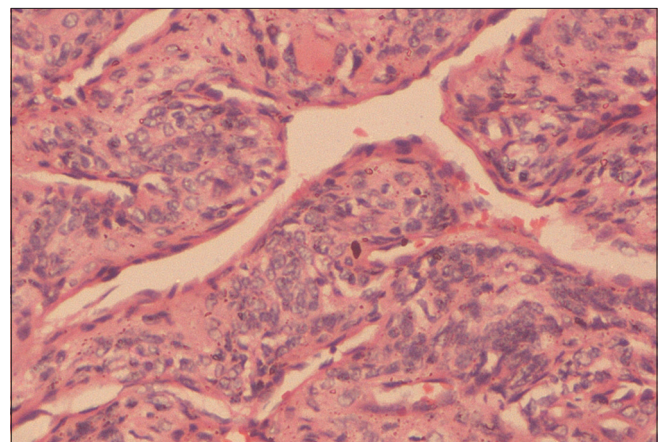


Figure 4: Typical staghorn pattern with proliferating lesional cells (H&E stain, x200)

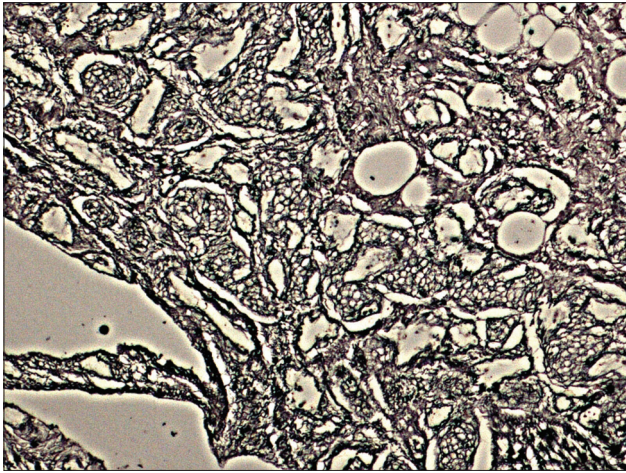


Figure 5: Reticulin stain showed distinctive reticulin pattern and each pericyte is surrounded by reticulin fibers (Reticulin stain, x100)

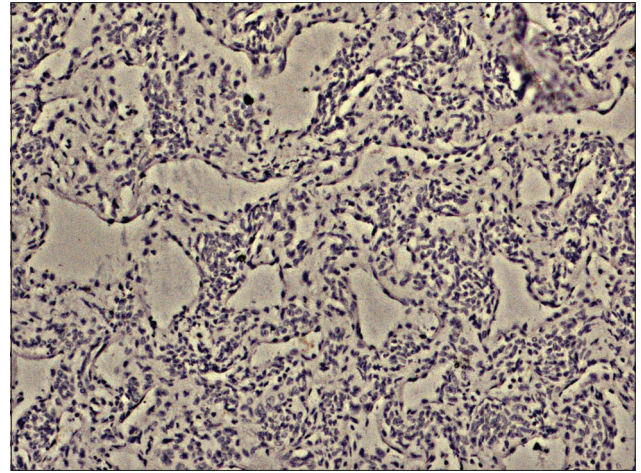


Figure 6: Immunohistochemistry revealing CD99 negativity (IHC stain, x100)

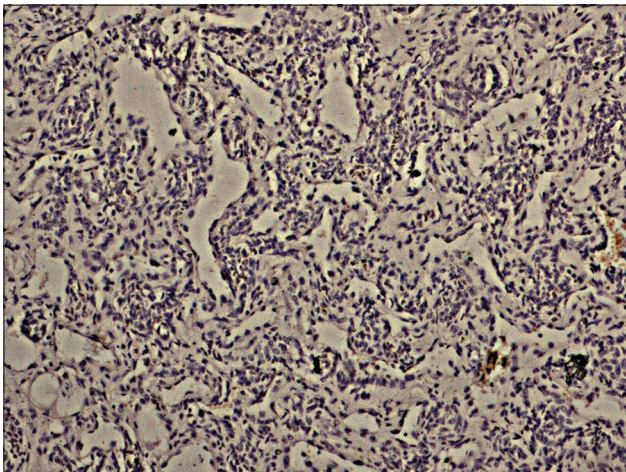


Figure 7: Immunohistochemistry revealing Bcl-2 negativity (IHC stain, x100)

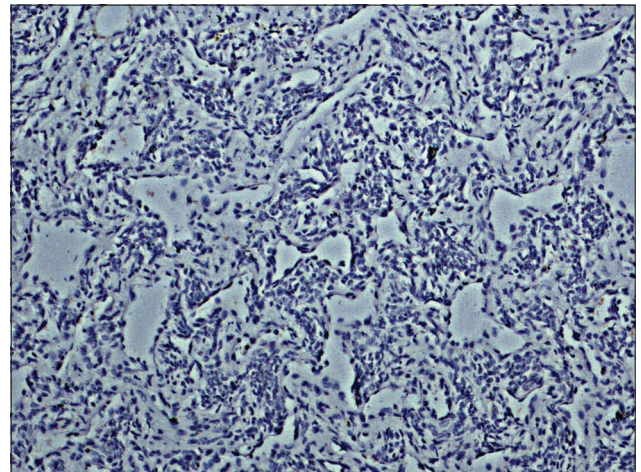


Figure 8: Immunohistochemistry revealing S-100 negativity with positivity in fat containing cells (IHC stain, x100)

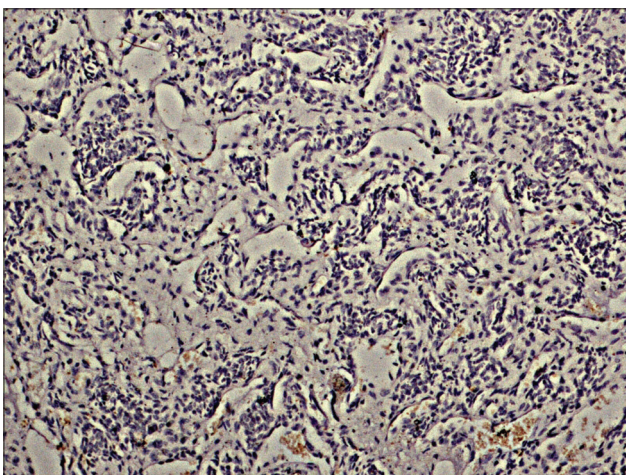


Figure 9: Immunohistochemistry revealing Pan cytokeratin negativity (IHC stain, x100)

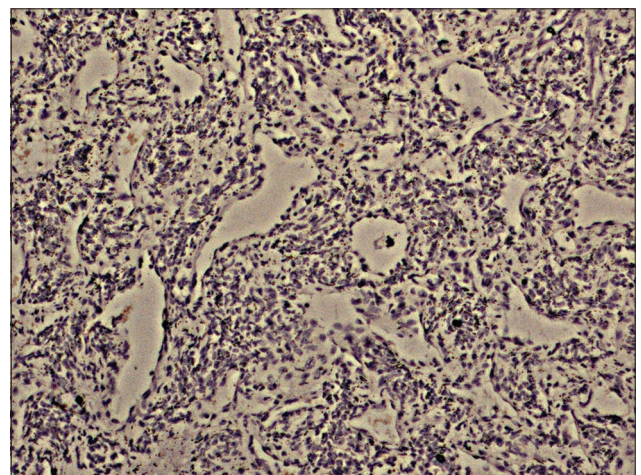


Figure 10: Immunohistochemistry revealing Desmin negativity (IHC stain, x100)

reported three cases of distinctive tumors composed of mature adipocytes and hemangiopericytomatous areas for which they

proposed the term LHPC. According to the WHO classification of soft-tissue tumors, hemangiopericytomas are closely related

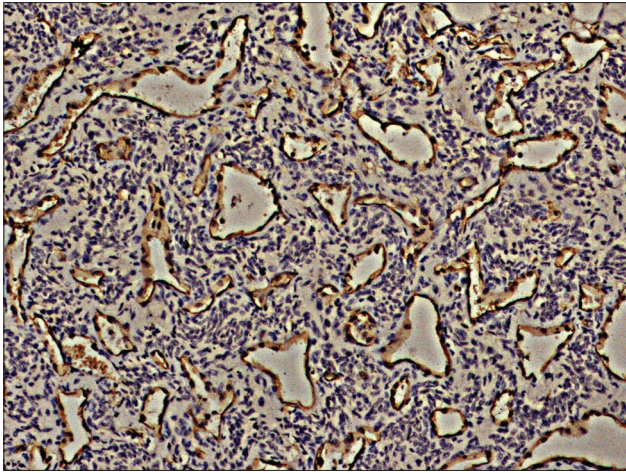


Figure 11: Immunohistochemistry revealing CD34 negativity in lesional cells, positivity in endothelial cells of vessels (IHC stain, x100)

to solitary fibrous tumors and are barely distinguishable as they share similar histological and ultrastructural features.^[3,4]

SFTs are subdivided into fibrous and cellular forms. Fibrous form is characterized by alternating hypercellular and hypocellular fibrous areas with many branching vessels with characteristically thickened and hyalinized walls. Cellular form is characterized by moderate to high cellularity with numerous thin walled vessels with little fibrosis and are indistinguishable from conventional HPC. In addition to these forms several unusual variants have been added like fat-forming variant called lipomatous hemangiopericytoma and giant cell angiofibroma which has giant multinucleated stromal cells.^[3]

Guillou *et al.*, suggested that LHPC does not correspond to a well-defined entity, but rather represents a fat-containing variant of SFT, since majority of the cases (but not all) show significant overlap histologically and immunohistochemically. Arguments in favor of this are: (1) Both are well-circumscribed, often nonencapsulated, superficial or deep-seated lesions (2) Both occur in middle-aged adults without apparent gender predilection (3) Both behave as indolent nonrecurring lesions in most cases (4) Both may contain a variable amount of mature fat cells, although this is rare in SFT (5) The great majority of L-HPC shows, at least focally, SFT morphology (6) Most L-HPC and SFT exhibit reactivity for CD34 and CD99 and, less frequently, for bcl-2 (7) L-HPC and SFT exhibit similar ultrastructural features.^[6]

Since its initial description, to date not more than 60 LHPC cases have been reported in the English literature. They occur in middle aged adults with an age range of 27–75 years, with a male predilection. They have a wide anatomical distribution including the pleura, mediastinum, thyroid, head and neck, but the retroperitoneum and deep soft tissues of the lower extremity, especially the thighs, are predominantly affected and involved in more than half of the affected cases. Clinically, most lesions present as a longstanding indolent tumor and do not metastasize or recur after a resection.

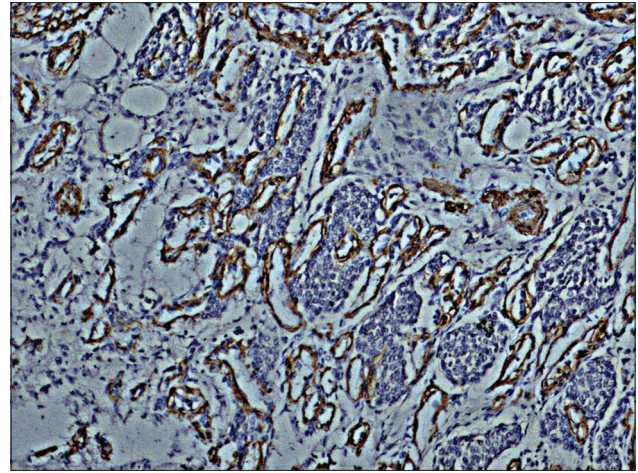


Figure 12: Immunohistochemistry revealing Smooth muscle actin negativity in lesional cells and positivity in vessel walls (arrow), (IHC stain, x100)

Conditions which mimic HPC clinically in the submandibular region include branchial cyst, dermoid cyst, plunging ranula, pleomorphic adenoma, arteriovenous hemangioma, deep seated lipoma, Hodgkin's lymphoma and non specific lymphadenopathy. Unlike the above conditions, HPC has no characteristic clinical features so the final diagnosis must depend solely on histopathology.

The histological appearance of HPC and SFT may be similar. However, differences between the two lesions exist. Although architectural variability is the hallmark of solitary fibrous tumor regardless of its location, the converse is true of hemangiopericytoma. Generally, HPC shows homogeneously higher cellularity and stag-horn shaped vessels throughout the lesion, whereas SFT shows varying cellularity and often thick and keloid-like hyalinization. Moreover, in SFT, the tumor cells are separated by linear rows of collagen, at least focally, a feature not found usually in HPC.^[7]

Main histological differential diagnoses include angiomylipoma, myolipoma or lipoleiomyoma, well-differentiated liposarcoma, low-grade dedifferentiated liposarcoma and spindle cell lipoma (SCL).^[3] Immunohistochemistry is very helpful in distinguishing these entities.

CD34 and CD99 may be found in almost every case and their detection has high diagnostic value.^[8] CD34, a haematopoietic progenitor cell membrane antigen, is of great diagnostic help in distinguishing SFT from HPC. SFTs are strongly positive for CD 34 whereas HPCs stain less frequently.^[9] CD34 points to a mesenchymal origin and are the most important marker to distinguish SFT from other spindle cell lesions. Strong CD34 immunoreactivity has also been observed in other tumors, like giant cell fibroblastoma, myofibroblastoma of the breast and in a high percentage of gastrointestinal stromal tumors. Thus, CD34 is not specific for SFT and needs to be considered in the appropriate histomorphologic context.^[10]

Recently it has been suggested that CD34 negativity could be associated with more aggressive behavior. The loss of CD34 might indicate a detachment of the tumor cell from the stem cell niche toward malignant transformation.^[11] Furthermore Wang *et al.*, suggested that loss of CD34 might promote tumor metastasis to other organs and could lead to malignant transformation from the benign tumor relevant to fatal outcome.^[12] Our present case was negative for CD34 and CD99 which we thought may be due to the above reasons. In the current literature there is only one case which showed negativity for both CD34 and CD99.^[13]

CD 99 and bcl-2 are the two markers which help in differentiating SFT from HPCs.^[14] According to Gengler *et al.*,^[3] and Mentzel *et al.*,^[15] the non adipocytic spindle cell component is invariably positive for CD 99 and less frequently for CD 34. Bcl-2 is also proved to be a marker which was consistently expressed in SFTs where as HPCs stain negative to bcl-2.^[16] However, reticulin stain can be used to confirm that the proliferating cells are outside the basement membrane of endothelial cells. HPC has a highly vascular pattern usually highlighted by a reticulin stain.

Despite great similarity between L-HPC and SFT, Guillou *et al.*, of the opinion that there are few L-HPC which shows little overlap with SFT and accordingly, might be kept in the ill-defined hemangiopericytoma category. Those lesions often show a well-developed “staghorn” branching vasculature, little fibrosis, little perivascular hyalinization and are CD34 and/or bcl-2 negative. They could represent the hemangiopericytoma end of the L-HPC spectrum.^[6]

Hemangiopericytoma with normal fat trapping also should be considered in the differential diagnosis. In our present case, fat was relatively present throughout the tumor (even in the center of the tumor surrounded by neoplastic cells). Furthermore, MRI of the present case also showed the presence of low intensity areas which are suggestive of fat in the tumor proper.

Despite negative staining for CD34 and CD99, the present case demonstrated consistent HPC vascular pattern throughout the tumor, with reticulin surrounding individual cells. So excluding other differential diagnostic considerations and taking in to consideration the reticulin pattern and histomorphology the diagnosis of lipomatous hemangiopericytoma was given.

In conclusion we report the first case of LHPC in a paediatric patient. Longterm follow-up is mandatory, as a subset of tumors with bland histology have been shown to behave aggressively. However, more number of cases of LHPC should be studied to provide further understanding of this rare tumor.

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How to cite this article: Madala J, Guttikonda VR, Korlepara R. Lipomatous hemangiopericytoma in a child: A case report with immunohistochemical evaluation. *J Oral Maxillofac Pathol* 2015;19:92-6.

Source of Support: Nil. **Conflict of Interest:** None declared.

A consent form has been received from the patient to reveal the identity of the patient