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CASE REPORT

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Glomangiomyoma: An unusual neck mass with atypical presentation in a child—A case report

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ABSTRACT

Background: Glomangiomyoma is a rare histological variant of glomus tumor (GT). Clinically, it was referred to as an enlarged cervical lymph node and is challenging to diagnose. Only one case has been reported in the neck of a child so far in the literature.
Case Summary: A 10-year-old girl presented with complaints of painful progressive neck swelling on the left side for two and a half years. Sonography and Contrast-enhanced Computed tomography neck suggested a slow flow vascular malformation and fine needle aspiration cytology was inconclusive, demonstrating only a few blood cells. The mass was removed in toto under general anesthesia without postoperative complications. The histopathology confirmed it to be glomangiomyoma
Conclusion: It is an extremely rare variant of GT and only one case of a Glomangiomyoma was reported in the neck of a child. Although it is a rare occurrence, when encountering a subcutaneous mass that initially appears as a venous malformation but exhibits pain, consideration should be given to GTs or related conditions.

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Introduction

The Glomus tumor (GT) is rare, composed of varying proportions of glomus cells, blood vessels, and smooth muscle cells, and accounts for approximately 1.6% of all soft tissue tumors [1]. GTs usually present in adults between the third and fourth decade of life. GTs are benign vascular tumors typically seen at the distal extremities which present as small nodules beneath the fingernail. There is an increased female predilication [2]. The patients present with symptoms of pain and cold hypersensitivity with localized tenderness. They usually present as small solitary tumors. GTs are classified as solid glomus tumors (25% of cases), glomangioma (60% of cases), and glomangiomyoma (15% of cases) based on the varying proportion of glomus cells, smooth cells, and blood vessels. We present a rare occurrence of Glomangiomyoma in the pediatric neck.

Case Report

A 10-year-old female child presented with gradually increasing swelling over the left side of the neck for the past 3 years. Now presented with mild localised pain came to our institution. Physical examination revealed a cervical mass and a diagnosis of cervical lymphadenopathy was made and referred by ear nose and throat (ENT) clinician to our department for ultrasound evaluation of the same. Ultrasound examination of the neck–both greyscale and Doppler studies showed a heteroechoic predominantly hypoechoic lesion with an indistinct border in the left level 2 region with few cystic areas and multiple tiny hyperechoic foci without shadowing diffusely scattered throughout the lesion with minimal internal vascularity.In view of multiple calcifications considering them as phleboliths a diagnosis of hemangioma was made (Fig. 1A and B). Fine needle aspiration cytology (FNAC) was done which showed only hemorrhage. We proceeded with computed tomography (CT) neck with contrast which revealed an ill-defined iso to hyperdense lesion with multiple specks of calcification in the left carotid space deep to sternocleidomastoid muscle infiltrating it. In contrast, the lesion shows minimal heterogeneous enhancement. The lesion was

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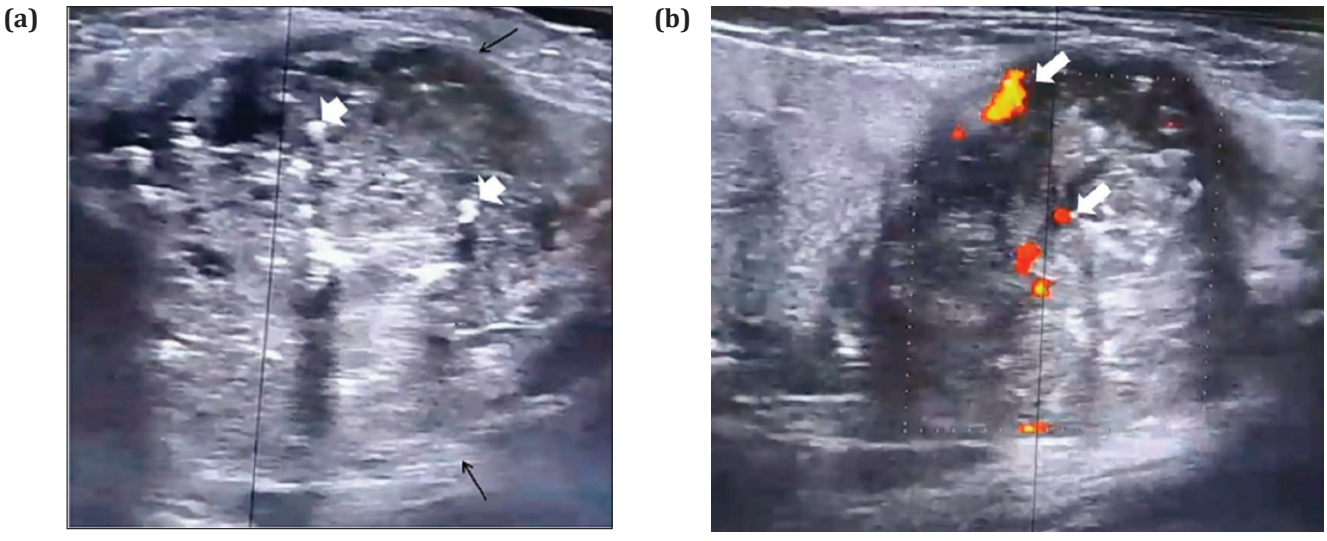


Figure 1. (a) Ultrasonogram of the left side of the neck showed a well-defined hypoechoic lesion (marked by black arrows) with few cystic areas and multiple punctate hyperechoic foci of calcification (marked by white arrows). (b) Doppler showed areas of internal and peripheral vascularity (marked by white arrows).

observed to insinuate between the external carotid artery (ECA) and internal carotid artery (ICA), causing splaying of the vessels without significant mass effect (Fig. 2). A final diagnosis of slow-flow venous malformation was given. ENT surgeon proceeded with an excision biopsy and sent for histopathological examination. Histopathology shows the presence of glomus cells and immunohistochemistry showed positive for CD31 and smooth muscle actin (SMA) which are markers for spindle-shaped muscle cells and blood vessels (Fig. 3). The final diagnosis of Glomangiomyoma was arrived at.

Discussion

GTs are usually benign perivascular neoplasms, most likely derived from modified smooth muscle cells of the neuromyoarterialglomus body. Glomangiomyoma is a rare variant of it. The Glomus body is a specialized form of arteriovenous anastomosis that regulates heat, located in the stratum reticularis of the dermis seen in the subungual region, the lateral areas of the digits, and the palm [2]. Extradigital sites include patella, chest wall, bone, stomach, colon, nerve, eyelid, nose, mediastinum, small bowel, rectum, urinary tract, lung, cervix, vagina, oral cavity, mesentery, heart, lymph nodes, larynx, back, and trachea. GTs usually present in the digits presenting in the middle-aged population with female predilection. Glomangiomyomas are a less frequent type accounting for 10% of all GTs and 20% of these cases are multiple. Symptoms include pain and gradually increasing swelling with sensitivity to temperature and touch. Previous literature shows neck Glomangiomyoma presenting as swelling with localized pain in the neck [3]. The differential diagnosis for a child presenting with long-standing painful swelling includes lymph node, venous anomaly, and nerve sheath tumors especially neuroblastoma in childhood. Cervical neuroblastoma are uncommon lateral neck mass lesion usually arising from the sympathetic ganglia. Imaging shows soft tissue density lesions with areas of calcification and increased vascularity. Radiological imaging is required to characterize the lesion and to know the extent of the lesion and its vascularity. Ultrasound is used to evaluate the solid nature of the lesion, calcification, and its vascularity. Contrast-enhanced CT provides additional information on the extent of the lesion, infiltrating of surrounding structures, and enhancement pattern. The treatment plan includes an excision biopsy of the lesion and confirm histopathological diagnosis.

On Ultrasound imaging, Glomangiomyoma appears as well well-defined hypoechoic lesion with multiple specks of calcification. Doppler showed minimal internal vascularity with a low resistive index. CT depicts iso to hyperdense lesion showing enhancement on contrast and splaying of vessels ensuring the site of glomus cells. Histologically, the tumor cells consist of varying proportions of glomus cells, vascular structures, and smooth muscle tissue. These are well-circumscribed lesions with tight convolutes of proliferating capillaries entangled by uniform glomus cells, the cohesive nature of the cells gives them an epithelioid appearance [4].

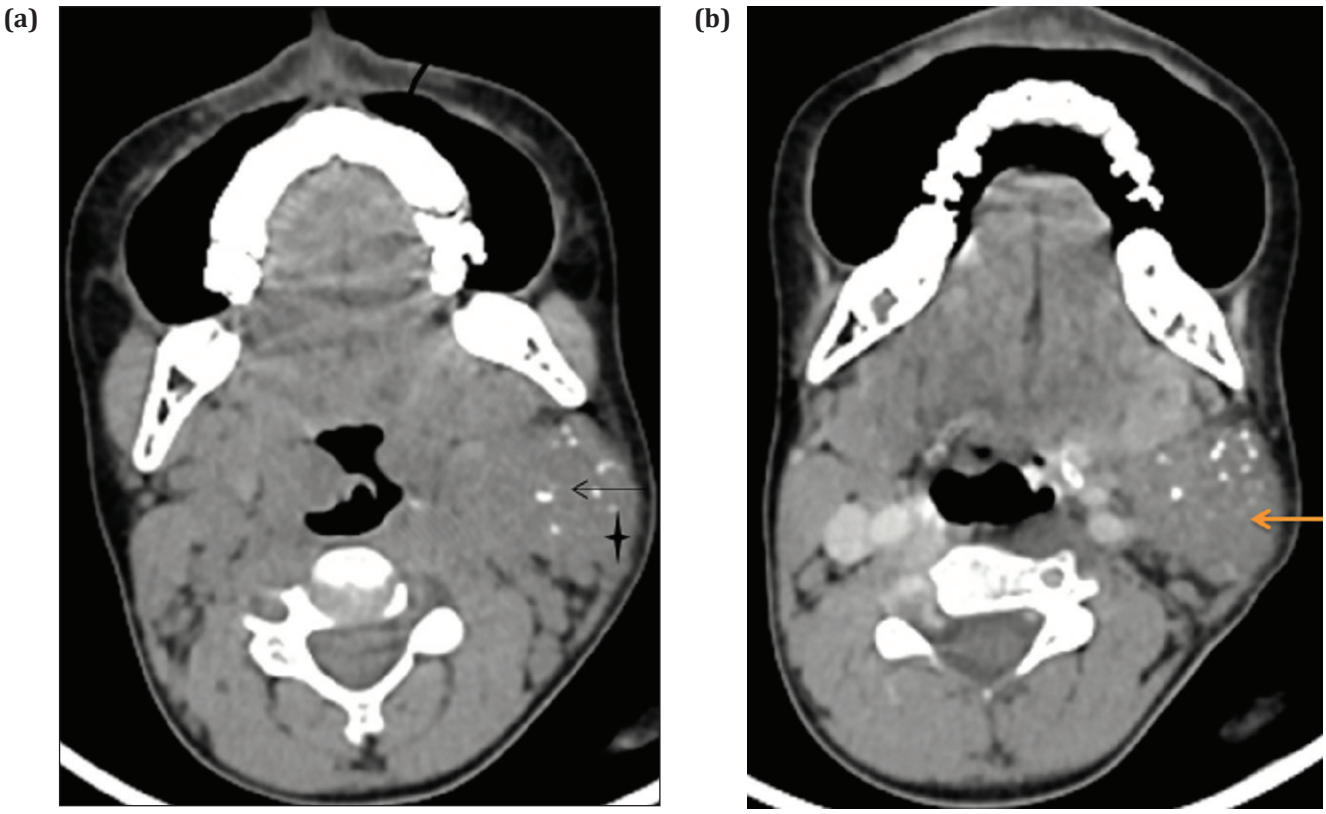


Figure 2. (a) Noncontrast CT axial section neck showed an isodense lesion with multiple calcific foci in left carotid space(marked by black arrow) lying deep to the sternocleidomastoid muscle (marked by "†") with loss of fat plane with the muscle. (b) Contrast CT axial section showed minimal enhancement of the lesion insinuating between the carotid vessels. (c) MIP images in the coronal section showed the splaying of ECA and ICA (marked by black arrows) by the lesion (marked by "†").

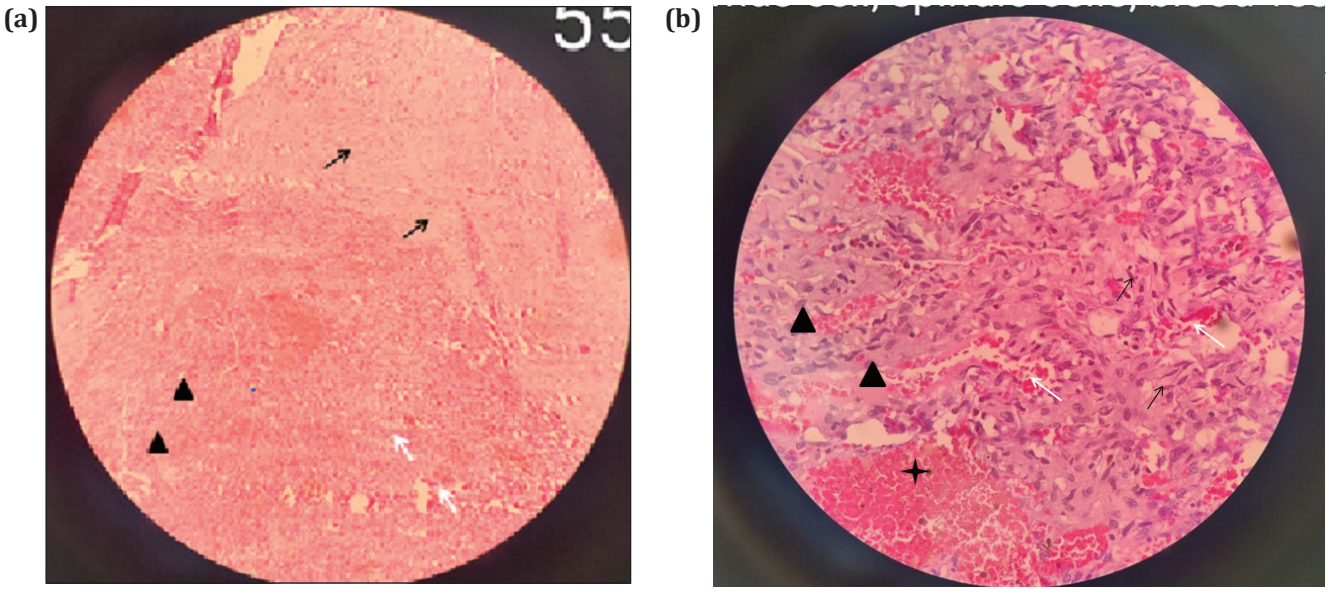


Figure 3. (a) Low power view showed prominent thin-walled blood vessels (marked by white arrow) with the proliferation of tumor cells (black arrow head) around it along with the variable proportion of smooth muscle(black arrow). (b) High power view showed prominent thin-walled blood vessels(marked by white arrow) with the proliferation of glomus cells (black arrow head) around it along with the variable proportion of smooth muscle(black arrow) and areas of calcification("†").

Glomangiomyomas resemble similar to that routine GT or a glomangioma. However, there are more elongated, mature smooth muscle cells. Immunohistochemically, all GTs show positive reactions for SMA and CD34. In our institution, the final diagnosis was arrived at with the help of imaging such as an ultrasonogram, contrast-enhanced CT, FNAC, and histopathological evaluation.

Surgical excision is the treatment of choice as it is a localized lesion in the head and neck. The child underwent an excision biopsy of the lesion. Intraoperatively it was a well-circumscribed lesion with no macroscopic signs of adjacent sternocleidomastoid muscle infiltration. There were no postoperative complications. Histopathological examination confirmed the diagnosis of left cervical Glomangiomyoma. Histopathology of surgical specimens showed no signs of atypia or mitotic activity. The child came for a review scan after 3 months which showed no residual lesion.

Conclusion

It is an extremely rare variant of GTs and only one case of a Glomangiomyoma is reported in the neck of a child. Although it is a rare occurrence, when encountering a subcutaneous mass that initially appears as a venous malformation but exhibits pain, consideration should be given to GTs or related conditions.

Consent for Publication

Written informed patient consent for publication has been obtained.

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