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Case Report

Glomangiopericytoma in a young girl: a rare case report and literature review

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Abstract

Glomangiopericytoma is a rare vascular neoplasm originating in modified perivascular glomus-like myoid cell. This tumor predominantly involves the sinonasal region but accounts for less than 0.5% of all sinonasal neoplasm. Majority of the patients presents in the sixth and seventh decade of life, the average being 50 years. A literature review of the last 10 years did not reveal any adolescent patient suffering from this type of neoplasm. A 15 years old girl presented with right nasal obstruction, endoscopic examination of the nose revealed a painless mass obstructing the right nasal cavity with normal nasal mucosa. CT and MRI scan revealed a right nasal cavity mass causing deviation of the nasal septum. Histological examination confirmed the diagnosis of glomangiopericytoma. It is important for the otolaryngologist to consider this type of neoplasm in adolescent to facilitate prompt diagnosis and treatment.

Keywords: Glomangiopericytoma, Sinonasal tumor, adolescent tumor, Paranasal Sinuses, Nasal Cavity

INTRODUCTION

Glomangiopericytoma is a rare vascular neoplasm that originates in modified perivascular glomus-like myoid cell and found mostly within the nasal cavity (Dandekar and McHugh, 2010). They are often termed sinonasal type-hemangiopericytoma and hemangiopericytoma, heman-giopericytoma-like tumor. But histopathologically they differ from hemangiopericytoma originating from other organs as they demonstrate more myogenic differen-tiation resembling a glomus tumor.

Sinonasal glomangiopericytoma is categorized into borderline and low-malignant-potential soft tissue tumors of the nose and paranasal sinuses (Dandekar and McHugh, 2010).

The term hemangiopericytoma was first described as a soft tissue tumor with a characteristic histological features by Stout and Murray in 1942 (Dandekar and McHugh, 2010). The sinonasal hemangiopericytoma was named glomangiopericytoma by The World Health Organization and was defined as a sinonasal tumour demonstrating perivascular myoid phenotype (Dandekar and McHugh, 2010). Also the tumor is thought to be derived from a modified perivascular glomus-like myoid cell (Dandekar and McHugh, 2010).

This tumor is mostly located within the nasal cavity itself, although some cases of paranasal sinuses involvement were reported (Lin et al., 2006).

Majority of the patients presents in the sixth and seventh decade of life (Thompson, 2004), the average being 50 years (Gillman and Pavlovich, 2004). Thorough literature review of the last 10 years did not yield any glomangiopericytoma patient in their adolescence. We report an extremely rare case of a 15 years old girl with a large glomangiopericytoma occupying the right nasal cavity presenting with a complaint of nasal obstruction.

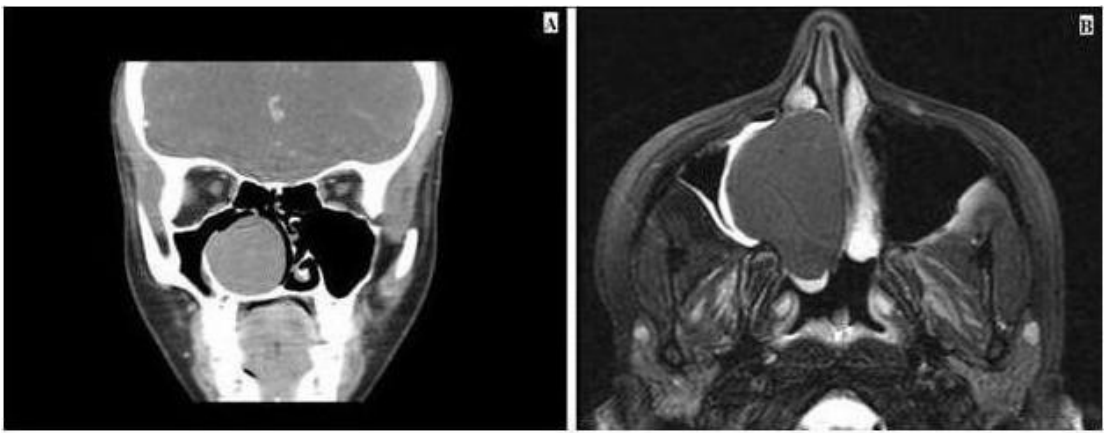

Figure 1. a) Coronal CT image shows a right sided nasal cavity mass caused obstruction and expansion of the right nasal cavity. **b)** Axial MRI image shows a homogeneous right nasal cavity mass extended to the postnasal space.

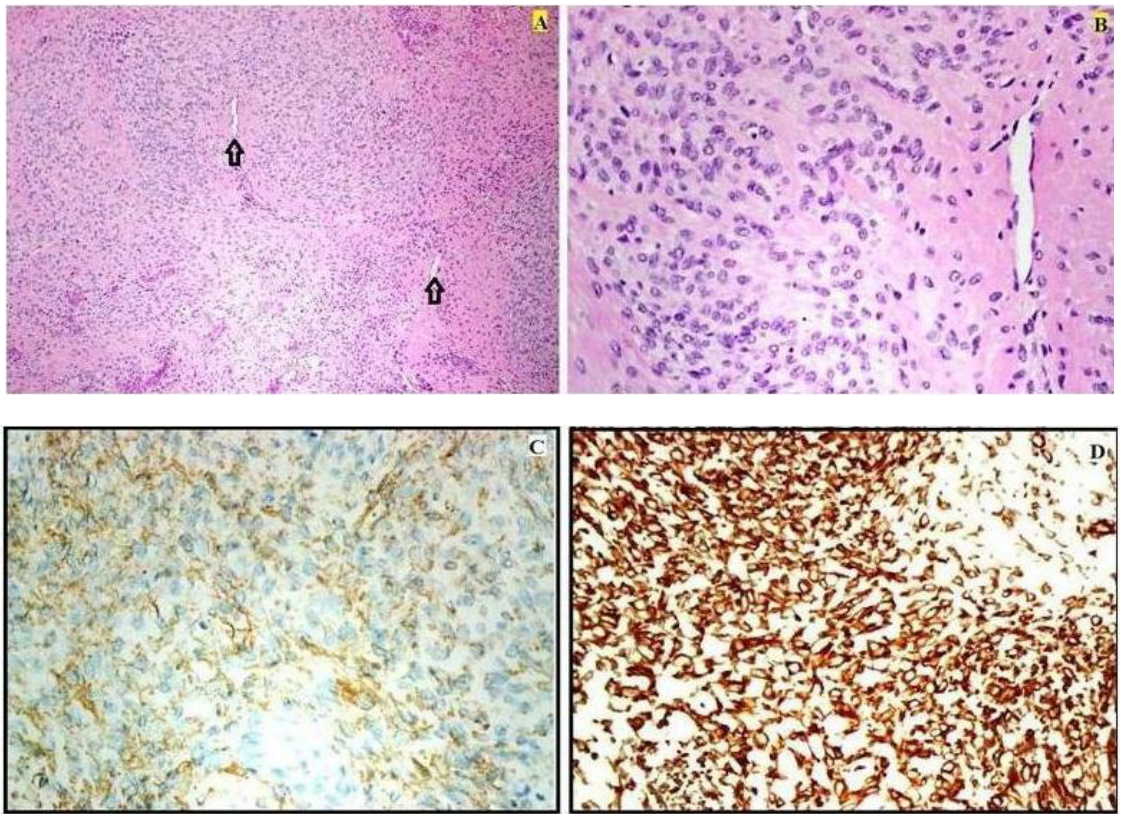

Figure 2. a) Moderately cellular spindle cell neoplasm with scattered thin-walled vessels (arrows) through the tumor (H/E stain, original magnification x 40). b) The neoplastic cells have indistinct cell borders with a small amount of eosinophilic cytoplasm. Nuclei have blunt ends and display mild atypia. Mitotic figures are infrequent. (H/E stain, original magnification x 400). Upon Immunohistochemically, the neoplastic cells are positive for smooth muscle actin (c) and vimentin (d). (Immunohistochemically stain, original magnification x 400).

CASE REPORT

A 15 years old female presented to our outpatient department complaining of right nasal obstruction. She denied any rhinorrhea, postnasal drip, epistaxis facial or nasal pain. Her medical history did not show any relevant

information regarding predisposing factors or any chronic problems. Examination using flexible nasolaryngoscope revealed a painless mass completely obscuring the right nasal cavity with normal nasal mucosa. CT scan and MRI of the paranasal sinuses were performed (Figure 1). CT scan revealed a 3.3 cm X 5 cm X 3.2 cm right sided nasal cavity mass causing expansion of the right nasal cavity with deviation of the nasal septum to the left. No destructive bony changes were observed. MRI scan revealed homogeneouslow signal intensity mass on T1 and T2-whighted sequences with homogeneous enhancement in the post contrast sequences. The mass was slightly extending posteriorly to the right side of the post nasal space. The mass did not invade surrounding structure. Endoscopic surgery was performed with full excision of the tumor.

Histopathology of the resected tumor (Figure 2) showed a moderately cellular tumor composed of spindle cells arranged mainly around blood vessels. No atypia were noticed and mitotic figures were infrequent. The neoplastic cells were positive for smooth muscle actin and vimentin and negative for desmin, CD34, CK, S-100 and EMA. The histological findings were compatible with glomangiopericytoma.

Patient recovered without any recurrence.

DISCUSSION

Glomangiopericytomas are rare and comprises less than 0.5% of all sinonasal neoplasms (Thompson, 2004). This tumor can affect any age however the incidence peaks in the sixth and seventh decade of life (Thompson, 2004).Our case is unique in the early age of presentation.

This tumor is affecting females more than males (Thompson, 2004; Higashi et al., 2011).More than sixty percent of patients with sinonasal hemangiopericytoma present with nasal obstruction and/or recurrent epistaxis. Additionally other nasal symptoms can include difficulty in breathing, rhinorrhea/postnasal drip, as well as extranasal symptoms like visual disturbance, pain, headache, serous otitis media and proptosis (Duval et al., 2013; Gillman and Pavlovich, 2004; Lin et al., 2006).Identified risk factors include hypertension, past trauma, corticosteroids use and pregnancy (Dandekar and McHugh, 2010; Wang and Chu, 2013). Clinically, this can be misdiagnosed as inflammatory polyp (Duval et al., 2013) as the tumor is polypoid, beefy red to grayish pink, edematous, fleshy to friable, soft and hemorrhagic (Thompson, 2004). Also a case is reported with metastatic lung cancer in remission, presented with epistaxis; it was identified as an isolated case of nasal glomangiopericytoma not related to primary disease (Higashi et al, 2011).

In histological examination, Hematoxylin and eosin stain shows subepithelial well delineated but non-encapsulated cellular tumor, it is comprised of closely packed cells and sometimes exhibiting a storiform, whorled or palisaded pattern, interspersed with numerous branching staghorn or antler-like configured vessels, the neoplastic cells are uniform with vesicular to hyperchromatic round to oval to spindle shaped nuclei. On the other hand, immunohistochemistry shows positive reaction for vimentin, muscle specific actin and α-smooth muscle actin, but negative for CD34 (Thompson, 2004; Dandekar and McHugh, 2010). Imaging studies are used to demonstrate the location, size and invasion to adjacent structure. MRI is preferred over CT scan in differentiating inflammatory fluid and tumor mass in obstructed paranasal sinuses (Szewczyk-Bieda et al., 2014; Ledderose et al., 2013). Other sinonasal tumors with histological features similar to glomangiopericytpma include solitary fibrous tumor, glomus tumor, leiomyoma, leiomyosarcoma, synovial sarcoma and haemangioma (Dandekar and McHugh, 2010; Dandekar and McHugh, 2010).

Total surgical excision with tumor free margins is the gold standard of treatment (Lin et al., 2006). Although this tumor is categorized as a borderline low malignancy tumor by WHO classification, cases of invasive behavior and distant metastasis have been reported (Dandekar and McHugh, 2010; Duval et al., 2013).The overall 5-year survival rate is excellent after complete removal. However, recurrence and tumor related death, although uncommon, have been reported (Duval et al., 2013; Higashi et al, 2011). It is believed that it might be a consequence of incomplete excision, making regular post-operative lifelong follow up necessary (Duval et al., 2013).

CONCLUSION

Glomangiopericytoma in adolescent age group is extremely rare. It should form part of the differential diagnosis adolescent patients presenting with nasal mass. Effective management entails total surgical excision. Contemplating the possibility of recurrence, a long-term clinical and endoscopic follow-up on one of the patients is recommended.

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Conflicts of Interest: None

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