Sinonasal Hemangiopericytoma In Adolescents: Histopathologic And Surgical Dilemma

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Citation

Abstract
Sinonasal Hemangiopericytoma (HP) is a rare vascular tumor believed to derive from perivascular modified smooth muscle cells (pericytes). It comprises only one percent of all vascular neoplasms and approximately three percent of all soft tissue sarcomas1, 2. The majority of HPs occur in the trunk and lower extremities, with the head and neck being involved in only about 7.5-16 percent of cases3, 4. The majority of the cases involving the head and neck occur in the nasal cavity, paranasal sinuses, orbit, parotid gland, and the neck5,6,7. These tumors occur in all age groups with no sex predilection, and a peak incidence between the 5th and 6th decade8. Angiofibroma is a soft tissue vascular tumor that usually occurs in the nasopharynx or less frequently, in the posterior nasal cavity in adolescent males25.

We report three cases of HPs occurring in adolescents, one in female & two in male’s. In males they were resembling as angiofibroma on clinical presentation & radiologically, whereas in female patient it was mimicking as a fibrous tumor involving nasopharynx and paranasal sinuses.

ABBREVIATIONS
hemangiopericytoma (HP)
Ear Nose Throat (ENT)
computed tomography (CT)

CASE REPORTS
CASE 1
A 16-year-old adolescent male presented to ENT (otolaryngology-Head Neck Surgery) outpatient services of Postgraduate Institute of Medical Education and Research, Chandigarh with one-year history of unilateral nasal obstruction and epistaxis. ENT examination showed a large purple mass in the left nasal fossa. The mass was smooth, soft and bled when touched. Contrast enhanced CT scan revealed well circumscribed, contrast-enhancing mass in left nasal cavity and ethmoid sinuses (figure1)

There was no bone destruction. Digital substraction angiography showed blood supply from ipsilateral internal maxillary artery only, due to this embolisation was not done. Provisional diagnosis of Juvenile Nasopharyngeal Angiofibroma was made. The patient was operated upon via Endoscopic Endonasal approach and the mass was completely removed. There was around 300ml of blood loss & this tumor was more vascular than was expected.

Figure 1
Figure 1: CT scan coronal cut showing contrast enhancing mass in posterior left nasal cavity and ethmoids.
Histology showed a low-grade hemangiopericytoma. The tumor cells were positive for vimentin. The tumor has not recurred during a three-year follow-up of this patient.

CASE 2
An 18-year-old adolescent male presented to the Emergency services of Postgraduate Institute of Medical Education and Research, Chandigarh with epistaxis. He gave two-year history of unilateral nasal obstruction and epistaxis. After initial management for epistaxis, the examination revealed dark red, mucosa-covered mass in nasal cavity and nasopharynx. The preliminary diagnosis of nasopharyngeal angiofibroma was made. Contrast enhanced CT scan revealed a contrast enhancing mass in nasal cavity and nasopharynx. Digital subtraction angiography confirmed the vascular nature of the tumor with the blood supply from internal maxillary artery. The patient was operated upon via lateral rhinotomy approach and the tumor was completely removed. Histopathological studies revealed the tumor to be nasopharyngeal angiofibroma. Six months postoperatively, patient was diagnosed for local recurrence. Fresh CT scan showed the tumor was involving nasopharynx, sphenoid sinus and pterygopalatine fossa with bony erosion (figures 2&3).

Figure 2
Figure 2: CT axial cut showing contrast enhancing mass in left nasopharynx and pterygopalatine fossa with erosion of the pterygoid plates.

The tumor was removed by Transantral approach using Weber Ferguson incision. This tumour was also not embolised and vascularity of this tumour seen intraoperatively was more. Histopathology revealed the tumor to be hemangiopericytoma. Previous slides of this patient were reviewed and diagnosis revised to hemangiopericytoma (figure4).

Figure 3
Figure 3: CT coronal cuts showing contrast enhancing mass in left nasopharynx and sphenoid sinus with erosion of root of pterygoid and intracranial extension

Figure 4
Figure 4: CT scan showing tumour in nasal cavity and going to anterior cranial fossa.

CASE 3
A 30 years old female patient with presenting complaint of
right-sided nasal obstruction of 6 months duration, nasal bleeding (intermittent, mild, spontaneous) with headache presented in the outpatient department. Examination revealed a pinkish fleshy mass, insensitive on touch, not bleeding on touch, and filling the nasal cavity. Biopsy was taken from this mass. Computed Tomography showed a mass filling the nasal cavity, ethmoid and sphenoid sinuses and going to the anterior cranial fossa. Biopsy revealed solitary tumour. The patient was planned for endoscopic clearance of the tumour. During surgery there was more bleeding but the tumour could be removed in piece meal. The histopathology revealed haemangiopericytoma. (Fig 5&6).

**Figure 5**
Figures 5 & 6: Histopathology.

**DISCUSSION**

Hemangiopericytoma (glomangiopericytoma) was first described by Stout and Murray in 1942, while studying the glomus tumor. About 150 cases have been reported in the English literature (Gudrun et al 1979; Abdel- Fattah et al 1990; Catalano et al 1996; Marianowski et al 1999). Most often, HP arise in the musculoskeletal system, the skin of the limbs, trunk and from the retroperitoneal area. In the Head and Neck, they usually develop in the soft tissues of scalp, face or neck. The tumor is twice as common in the nasal cavity as in the Para nasal sinuses. The sphenoid and ethmoid sinuses are involved four times more often than the maxillary sinuses.

The etiology of HPs remains unknown. Previous history of trauma has raised the possibility that this may stimulate proliferation of pericytes following damage to the capillary network and long term steroid therapy and arterial hypertension have been suggested, although none of these theories have been proved.

Most affected patients experience nasal obstruction and epistaxis along with a wide array of other nonspecific findings that are generally present for less than one year. Their natural history is uncertain but they are generally slow growing, solitary and indolent with local infiltration.

Histologically, these tumors are submucosal, usually covered by an intact respiratory epithelium. There is a diffuse growth of closely packed cells that appear in short interlacing fascicles (storiform, whorled, and palisaded patterns can be seen) that are richly vascularized. The vascular channels
range from capillary size to large patulous spaces that may have a ramifying “staghorn” or “antler-like” configuration. The neoplastic cells form a closely packed syncytium of uniform, monotonous, oval to slightly spindle-shaped cells with indistinct cell borders that contain vesicular to hyperchromatic, round to oval to spindle shaped nuclei. The tumor cells are immunoreactive with actins and vimentin but not with CD34, CD31, or FVIII-R Ag. A “hemangiopericytoma-like” pattern can be found in a wide array of neoplasms of divergent differentiation (eg. lobular capillary hemangiomas, angiofibromas, meningiomas, and leiomyomas), but the characteristic histologic and immunophenotypic features allow for separation.

Several authors have addressed the grading of HP as benign, borderline, and low-grade malignant, and overtly malignant. The mitotic rate has been used to distinguish between these categories, with lesions demonstrating over four mitoses per high power microscopic field being defined as malignant. Although these categories exist, the natural history of the disease is not correlated with the histological grading and is rather unpredictable, even if the tumor is benign histologically. A large tumor size (>6.5cm) has been correlated with poorer outcomes. Distant metastases occur in 20-50 percent of HPs involving all body sites, and in about 10 percent of cases involving the head and neck only.

The treatment of choice is wide surgical excision, due to its latent potential for malignancy. In parasanal sinus localizations, an external procedure is most frequently chosen to achieve total tumor excision, but when the tumor is purely intranasal or strictly located in the ethmoid or sphenoid sinus, it can be removed via an endoscopic endonasal approach. The use of radiotherapy and chemotherapy as adjuvant treatments is still controversial. Radiotherapy alone is reported to have a cure rate of only 13.3 percent.

CONCLUSION

HPs are uncommon vascular tumors, rarely located in the nasal cavity and paranasal sinuses. When they present in adolescent age group, they are difficult to distinguish from nasopharyngeal angiofibromas as was seen in our patients, i.e. Clinically, Radiological and even Histopathologically. Clinically they present as pinkish nasal masses, Radiologically HP shows some bony destruction as compared to angiofibromas that points some suspicion on diagnosis of angiofibromas preoperatively, Surgically HP are more vascular than angiofibromas and other nasal & sinus tumors, so requires a faster resection. Most importantly HP are important to be identified histopathologically as in all of our three cases these were diagnosed as some other tumour and it was only after reviewing the slides again and after proper staining final diagnosis of HP could be made. Surgical removal is the treatment of choice and tumor excision must be complete due to the latent potential for malignant behavior of these tumors.

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