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Haemangiopericytoma - A Rare Sino-Nasal Mass

Ruta Shanmugam Vellavedu Umapathy Shanmugam Rajagopalan Mariappan Balaji Swaminathan Srikanth Nandipati Dony Manattu Mathaikutty

Annamalai University,Chidambaram, India

Abstract

Haemangiopericytoma is a rare vascular tumor accounting for about 1% of vascular tumors of the body. Less than 5% of hemangiopericytoma occur in the nasal cavity and are characterized by a rather benign nature with low tendency of metastasis. We report a case of right nasal polyp which underwent treatment but histologically proved to be haemangiopericytoma.

Key-words: Haemangiopericytoma, sino-nasal, rare

Introduction

Haemangiopericytoma is a rare vascular tumor arising by the proliferation of pericytes.[1-3] They can arise anywhere in the body but an appreciable number (15-20%) occur in head and neck particularly in the soft tissues of scalp, face & neck. An origin in the oral cavity, nasal cavity or paranasal sinuses is less common.[4-6] The tumor may develop over a period of months and even years and presents in patients of all ages.[7] However, due to its rare occurrence, unpredictable biological behavior and unknown natural history, this entity usually triggers some confusion and some uncertainty about its management.[3]

Case report

A nine year old female child presented to the department of otolaryngology with right sided nasal obstruction and epistaxis. On clinical examination, a reddish polypoidal mass arising in the lateral wall of the right nasal cavity which was insensitive to touch, soft in consistency, bled on touch and was well circumscribed and it resembled an infected nasal polyp.

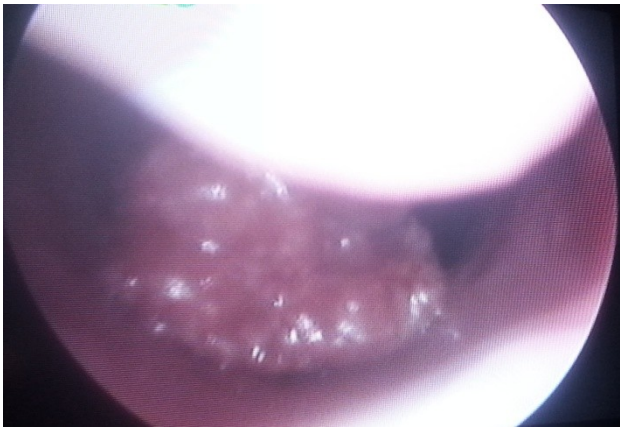

Fig 1. Diagnostic nasal endoscopy

CT scan showed the mass to arise from the right maxillary sinus extending into the nasal cavity, with mucosal opacity in the right frontal and ethmoid sinuses. No bony erosion was found (figure 2).

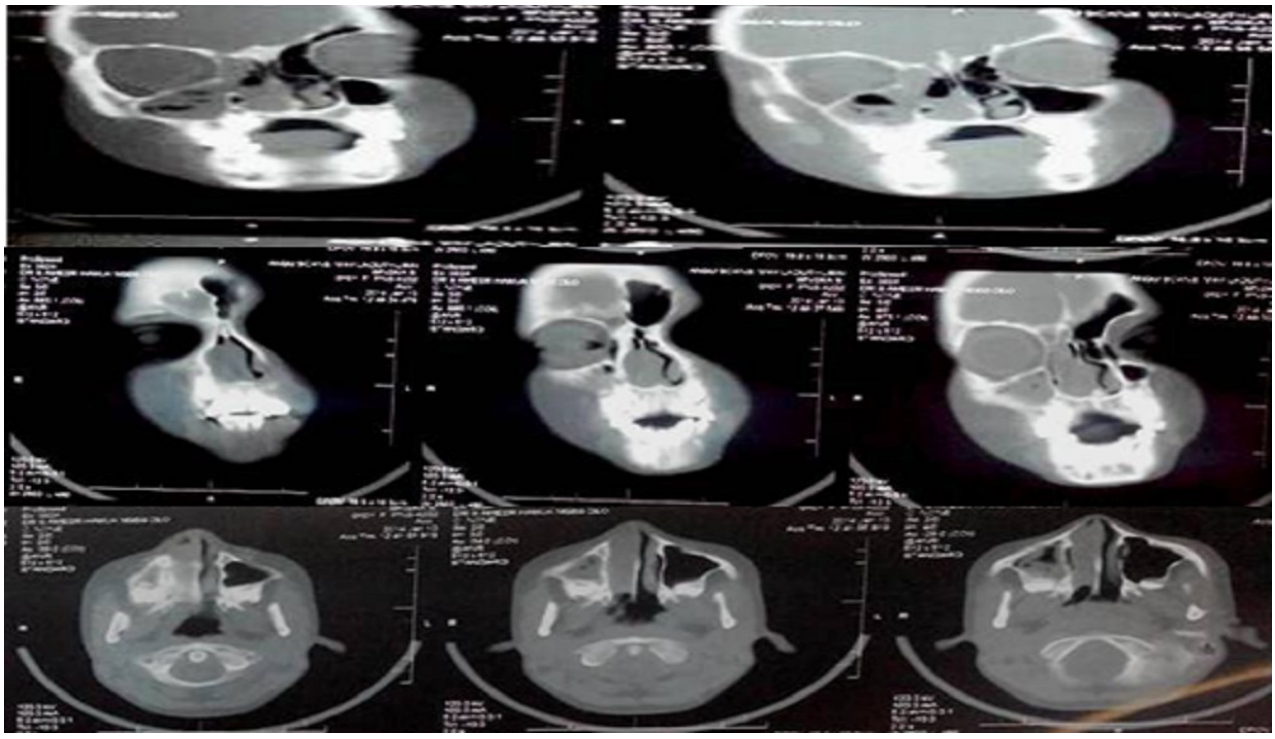

Fig 2. CT scan PNS

Patient underwent endoscopic guided polypectomy during which there was profuse bleeding for which the entire pedicle was cauterized and bilateral anterior nasal packing was done and the specimen was sent for histopathological examination. On pack removal, profuse bleeding was encountered and repeat electro cauterization with bilateral anterior nasal packing was done. Repeated bleeding from the site made us to revisit our clinical diagnosis and to our surprise it turned out to be haemangiopericytoma on histolopathology.

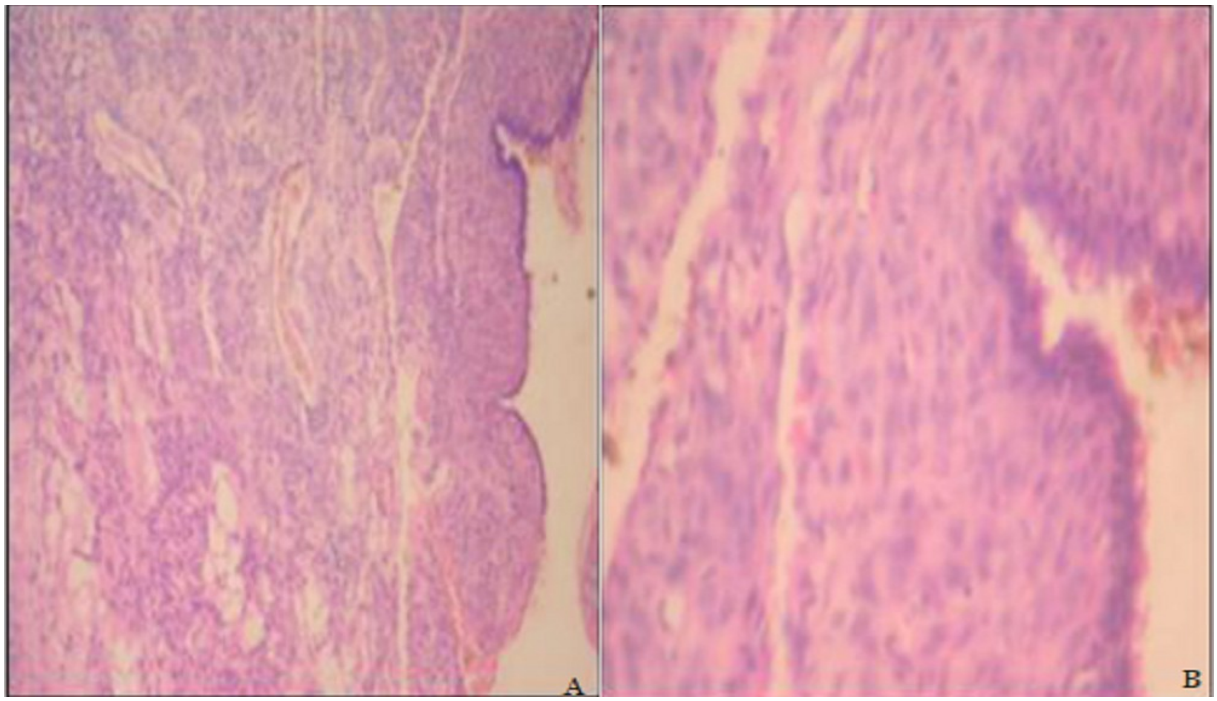

Fig 3. A (Low power view), **B** (High power view), Respiratory epithelium with tumor composed of vascular channels deeper to it.

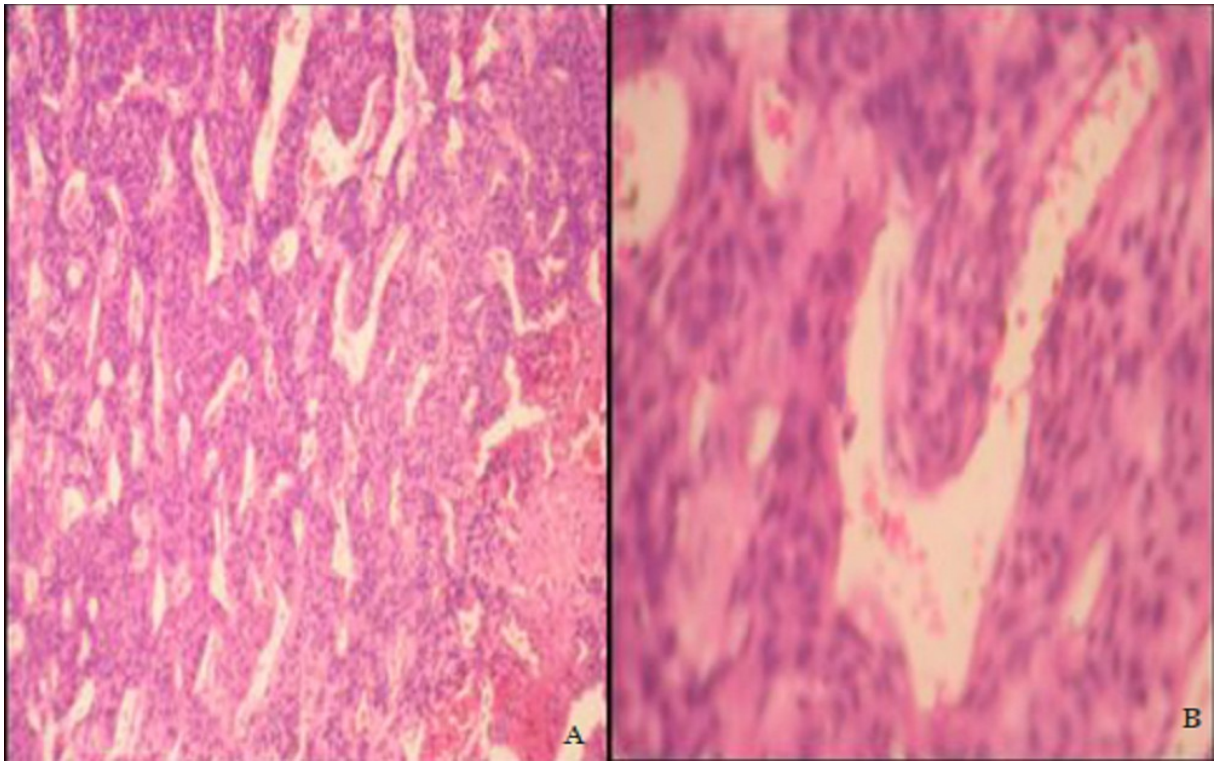

Fig 4. A (Low power view), **B** (High power view), Multiple vascular channels showing staghorn pattern lined by endothelial cells and lacking muscular coat surrounded by pericytes which are spindle shaped

Postoperative period was uneventful. She has been followed up for six months and no recurrence was found.

Discussion

The term pericyte was introduced by Zimmermann [2-5] in 1923 to describe cells encircling the blood vessel wall, consisting of thin processes and a prominent nucleus embedded in the basement membrane surrounding the capillaries. Stout and Murray [8-9] in 1942 coined the term "hemangiopericytoma".

Sino-nasal haemangiopericytoma usually follows a benign course, unlike those that arises in other somatic sites. Behavior is generally indolent though very rare metastatic tumors have been described. Sino-nasal hemangiopericytoma although classified as malignant neoplasm they can clinically manifest as either benign or malignant.[9] Local recurrence is seen in 25% and metastasis in 5% of the cases. Metastasis and recurrences have been described even decades after the first tumor treatment, emphasizing the need for lifelong follow-up. Metastasis occurs most commonly to lung and skeleton followed by chest wall, brain, bowel, orbit, lymph node.[1,3,5]

Haemangiopericytoma affect all ages, majority presenting between the 20^{th} and 70^{th} decade (median age 46 years). Incidence in childhood has been reported to be about 10%. Indeed, very few cases of pediatric head and neck haemangiopericytoma have been documented. The youngest documented age is of a two and a half year old. Gender or race predominance is not well demonstrated.[9]

Clinically, they are characterized as slowly expanding solitary mass that range in size from 1 cm to 20 cm. Intranasal haemangiopericytomas manifest as pale, grey white, well circumscribed mass with a soft, rubbery consistency resembling nasal polyp. The discoloration is explained by the compression of capillary lumen by proliferating pericytes. They are usually painless unless confined to an unyielding space. Nasal obstructions with epistaxis are the most common symptoms.[3,5,10] Vision impairment, headache and local swelling are less frequent symptoms as well.[10]

On ENT examination, sinonasal hemangiopericytoma is frequently mistaken for inflammatory polyp although, only histopathology can confirm the final diagnosis.[10] Grossly, they are lobulated or nodular, have well defined capsule, firmly attached to muscle or fascia. Sometimes they can be soft, spongy, firm or friable. Sizes greater than 6.5 cm; four or five mitotic figures per ten high power fields, cellular anaplasia are known to be associated with aggressive behavior.[3,5,7,9]

Microscopically, the tumor shows small to medium sized vascular channels lined by inconspicuous to flattened endothelial cells surrounded by spindle-shaped to oval, plump cells arranged radially and in sheets. Silver reticulum stain outlines the basement membrane and demonstrates the extra vascular position of the tumor cells. This stain is essential to establish the diagnosis of haemangiopericytoma.[3]

The treatment although controversial, most authors recommend wide local excision.[1,3,5,9,10] Lateral rhinotomy has been the most frequently used surgical approach, although endoscopic techniques may be appropriate in selected cases.[2,10] Jon K. Thiringer et al[2] suggested a conservative approach which should consist of debulking the tumor so that sinonasal obstruction is relieved and careful follow-up with serial endoscopic examinations and imaging studies. They recommend aggressive resection, only if the tumor shows a propensity for growth and local invasion. Radiotherapy and chemotherapy have limited use in these tumors.[2,8] These patients require lifelong follow-up because of their propensity for local and distant recurrence of the disease.[3,5]

Conclusion

Whenever a painless nasal soft tissue mass is diagnosed, one should include haemangiopericytoma in the differential diagnosis.[2-3] Misdiagnosis can lead to profuse bleeding during surgery which can be controlled by electrocautery based on our experience. Only excision biopsy of the lesion can confirm the diagnosis of haemangiopericytoma. Haemangiopericytomas are often difficult to eradicate and have been reported to recur even twenty years after the initial treatment. Thus, careful follow-up is the norm.[3]

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