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Hemangiopericytoma of the nasal cavity

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SUMMARY

Hemangiopericytomas are unusual vascular tumours that rarely occur in the paranasal sinuses and nasal cavity. They are thought to arise from pericytes that surround capillaries, however, there is no proven etiology of these tumours. The diagnosis cannot be made based on gross morphologic characteristics nor on frozen section biopsy. Instead, the diagnosis is dependent on careful histologic examination and reticulum staining. The diagnosis can be confirmed by electron microscopy. The histologic picture is greatly varied from tumour to tumour and within a given tumour itself.

The clinical course of hemangiopericytomas ranges from benign to malignant with possible distant metastases and cannot be reliably predicted by histologic criteria. Hemangiopericytomas of the nose and paranasal sinuses are thought to behave less aggressively than those occurring in other parts of the body. Lymph node metastasis is rare and elective neck dissection is not indicated.

The accepted treatment of these tumours is wide surgical excision, however adequate surgical margins are usually difficult in the sinonasal region. As a result, hemangiopericytomas can exhibit a high recurrence rate. Therefore, it is mandatory that these patients be followed carefully for the remainder of their lives.

INTRODUCTION

Stout and Murray are credited with describing hemangiopericytoma as a distinct form of vascular tumour in 1942. The tumour can arise from any place where capillaries are found and is thought to originate from pericytes (Compagno, 1978; Batsakis, 1979, 1983; Gudrun, 1979).

Pericytes are round or spindle shaped cells with long processes that are applied to the outer wall of the venous capillaries and post-capillary venules. They are completely surrounded by basement membrane and are thought to give mechanical

support to the capillary. They have contractile properties, and regulate the lumen size (Gudrun, 1979; Anderson, 1984).

The diagnosis of hemangiopericytoma can only be made by proper histopathological examination using special staining techniques. The diagnosis cannot be made on the basis of clinical, gross, macroscopic or fine needle aspiration findings (Compagno, 1978; Batsakis, 1979; Gudrun, 1979; Nguyen and Neifer, 1985). The majority of hemangiopericytomas are benign, however, malignant hemangiopericytomas have been described (Compagno, 1978; Gudrun, 1979; Nguyen and Neifer, 1985).

There have been approximately 23 cases of hemangiopericytomas involving the nasal cavity and paranasal sinuses reported to date (Eneroth et al., 1970). The purpose of this paper is to add to the literature another case of a hemangiopericytoma involving the nasal cavity and present a review of the pertinent literature.

CASE REPORT

A 29 year old white male had been treated for left sided nasal obstruction for approximately nine months. He failed to improve despite therapy and was referred to the Otolaryngology service at Mount Sinai Hospital in Chicago for evaluation. He complained of nasal obstruction and a slowly developing left facial deformity, but denied epistaxis or visual complaints. His medical history was unremarkable except for a previous orchiopexy.

Examination revealed a 3 by 4 cm area of swelling externally on the left side of the nose and a mass occluding the left nostril. The pupils were normal as were the eye movements. The oral cavity and pharynx were normal. There were no middle ear effusions noted and there were no palpable adenopathies in the parotid region or in the neck.

Plain radiographs of the paranasal sinuses showed opacification of the left maxillary sinus and nasal cavity. Tomograms of the paranasal sinuses and nasopharynx revealed a mass occupying the left nasal cavity which extended into the left maxillary sinus and to the nasopharynx. Bilateral carotid arteriogram demonstrated a hypervascular tumour in the left nasal cavity with its main blood supply originating from the left sphenopalatine artery, although additional blood supply was derived from the septal and alar branches of the superior labial artery and from the left anterior and posterior ethmoidal arteries. There was also significant cross circulation from the right sphenopalatine artery. A chest radiograph was normal.

The lesion was biopsied under local anaesthesia with a moderate amount of bleeding. The histopathological diagnosis was nasal hemangioma. Following ligation of the external carotid artery, the tumour was removed through a left lateral rhinotomy. Excision required a partial maxillectomy and complete ethmoidectomy. The estimated operative blood loss was 2000 cc. The patient did

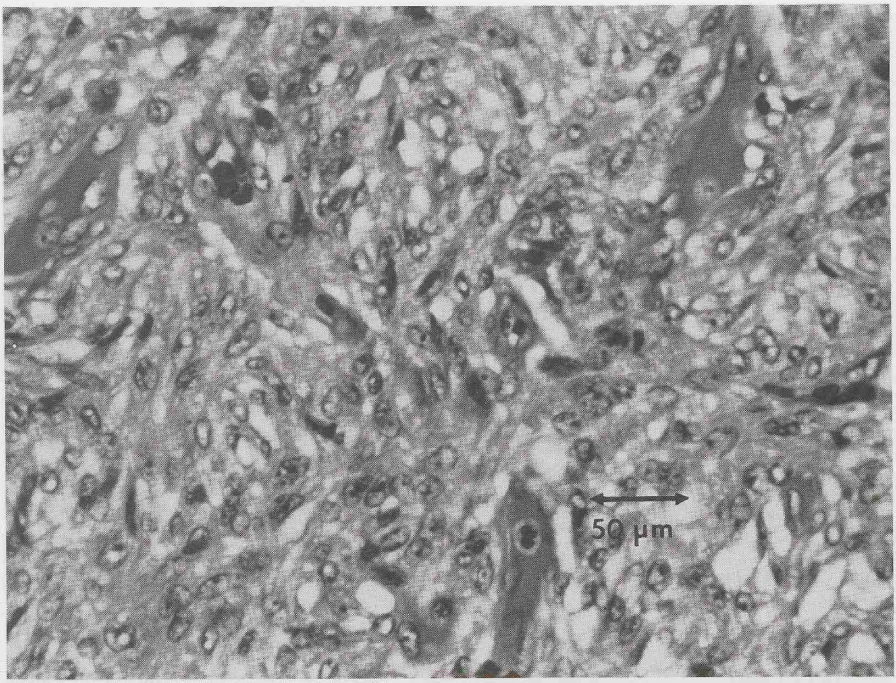

Figure 1. The tumour consists of fusiform spindle cells with frequent capillaries. (H & E stain, ×250)

well post-operatively and was discharged eight days later in good condition. Five years later he had no evidence of recurrence.

Histological examination of the surgical specimen demonstrated a richly vascularized tumour composed of endothelial channels surrounded by a densely packed stroma made up of round to ovoid cells or fusiform cells (Figure 1). The cells exhibited minimal pleomorphism, however, in several sections there were multiple mitoses. On reticulin stain, the tumour cells were found to be embedded in a rich mesh of reticulin fibers (Figure 2). Electron microscopy revealed mesenchymal cell features with the conspicuous peripheral basal lamina material encasing tumour cells, typical of pericytes (Figure 3). Due to the frequent mitoses in some regions, the tumour was considered to be a low grade malignancy.

DISCUSSION

Hemangiopericytomas are among the least common tumours of the head and neck, accounting for approximately 1% of vascular tumours (Batsakis et al., 1983). The most common areas for these tumours to arise from is in the musculoskeletal system and skin of the limbs and trunk and the retroperitoneal area (Batsakis,

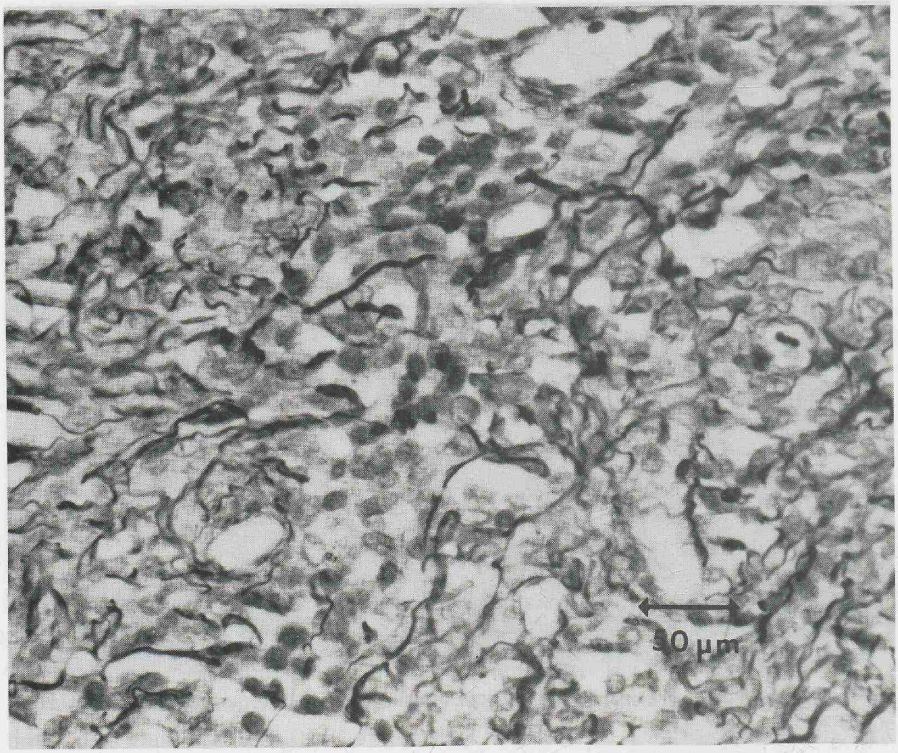

Figure 2. The tumour cells are extracapillary and embedded in a rich meshwork of reticulin fibres. (Reticulin stain, ×250)

1979). Approximately 15% to 25% of all hemangiopericytomas occur in the head and neck (Batsakis, 1979; Blitzer et al., 1985). In this region they usually arise in the soft tissues of the scalp, face or neck (Batsakis et al., 1983; Blitzer et al., 1985). Only 5% of all hemangiopericytomas occur in the nose and paranasal sinuses (McMaster et al., 1975; Batsakis, 1981). Occurrence in the nasal cavity is about twice as common as in the paranasal sinuses. The sphenoid and ethmoid sinuses are involved four times more often than the maxillary sinus (Batsakis, 1979; Blitzer et al., 1985). There appears to be no predilection for either sex or any age group; although there is a slight peak incidence occurring in the 5th an 6th decades of life (Gudrun, 1979; Anderson, 1984).

The true etiology of hemangiopericytomas remains unknown. Previous trauma has been implicated in the etiology of hemangiopericytomas. In a series of hemangiopericytomas presented by Stout, six of 31 patients gave a history of ecchymosis or swelling that did not improve as an initial sign of the tumour. It was thought that damage to the capillaries stimulated the proliferation of peri-

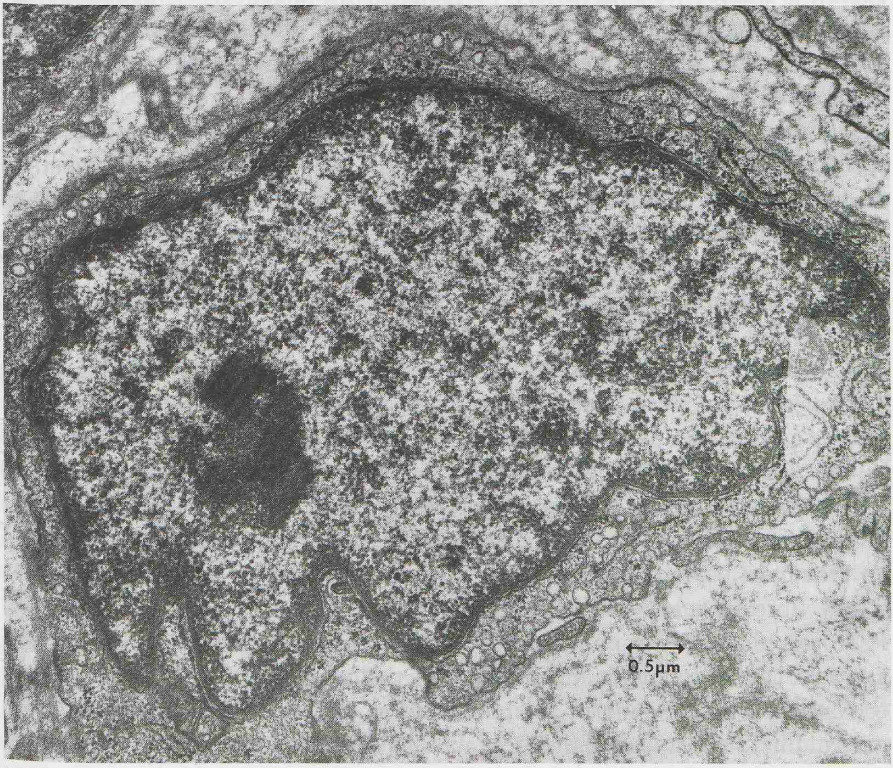

Figure 3. An electron micrograph of a representative tumour cell showing the periphery of the cytoplasm surrounded by a basal lamina. The cytoplasm is slightly fibrillar with rare profiles of endoplasmic reticulum and pinocytotic vesicles. (Uranyl acetate and lead citrate, ×24,000, courtesy of Pathology department at Mt. Sinai Hospital, Chicago IL.)

cytes. Long-term steroid therapy has also been suggested as an etiologic factor of hemangiopericytomas. However, none of these theories has been proven (Gudrun, 1979; De Campora et al., 1983).

Clinically, hemangiopericytomas of the nasal cavity are similar to other tumours of this region. They commonly present with persistent unilateral nasal obstruction and mild recurrent epistaxis. These tumours are usually painless, slow growing masses that are soft, polypoid and gray or red in colour. Extensive tumours may cause an external deformity of the maxilla.

Fine needle aspiration has not been found to be helpful in diagnosing hemangiopericytomas, or in discerning their benign or malignant nature (Nguyen and Neifer, 1985). The diagnosis of a hemangiopericytoma is difficult and is dependent on histologic examination with special staining techniques. In our patient the original biopsy was interpreted as a hemangioma and additional histo-

chemical studies were necessary to establish the diagnosis of hemangiopericytoma. Hematoxylin and Eosin stain can be used to identify the tumour, but the silver reticulin stain is necessary to confirm the diagnosis (Batsakis, 1979; Gudrun, 1979; De Campora et al., 1983). This stain demonstrates the cell-vessel relationship, making the sheathes of capillaries appear black and easy to recognize. It also demonstrates that the tumour is actually outside the sheath, a finding that helps differentiate it from other tumours such as a hemangioendothelioma (Batsakis, 1979; Gudrun, 1979). Electron microscopy can demonstrate the characteristic features of the pericytes, namely basal lamina, fibrillar cytoplasmic matrix, and peripheral cytoplasmic densities. In fact, Batsakis et al. (1983) used electron microscopy to confirm that the tumour cells of hemangiopericytomas originated from the pericytes of capillaries. Histologically, hemangiopericytomas exhibit a dense vascular bed that on light microscopy creates a staghorn appearance. The stroma of the tumour is made of round to spindle shaped cells with indistinct margins with oval or round nuclei. And at times, the stroma is so densely packed that the vascular channels appear to be obliterated (Figure 1). The stroma may exhibit a storiform pattern that resembles a fibrous histiocytoma. There may be varying amounts of cellular and nuclear atypia, fibrosis, haemorrhage, or necrosis.

Radiographic examination is important to define the extent of the tumour and allow proper treatment planning. Plain film radiographs of the sinuses and skull, conventional tomography, or computed tomography can be used to demonstrate the tumour limits as well as the presence of bony destruction, intracranial extension or extension into the orbit. Arteriography is useful to identify the main blood supply of the tumour. There have been no studies to determine if preoperative embolization of hemangiopericytomas of the nasal cavity is beneficial. Hemangiopericytonas of the head and neck have an unpredictable behaviour. Batsakis and Rice (1981) suggest that hemangiopericytomas of the nasal cavity and paranasal sinuses may have a less aggressive course than hemangiopericytomas of other sites. Hemangiopericytomas in general have been divided into benign and malignant categories based on factors such as the number of mitoses, cellularity, size, site of origin, and degree of anaplasia. Some authors claim that tumours with high numbers of mitoses i.e.4 per 10 high power fields, are associated with decreased survival (Enzinger and Smith, 1976). Large tumours, i.e. 6.5 cm have been described as being more aggressive with a 10 year survival of 63% compared to 92% 10 year survival of tumours measuring less than 6.5 cm (Enzinger and Smith, 1976; Batsakis and Rice, 1981). Hemangiopericytomas with large areas of necrosis may also be associated with a poorer prognosis (Blitzer et al., 1985). Tumour recurrence is also associated with a poorer prognosis. Metastases were seen in 69% of recurrent tumours according to Enzinger and Smith (1976).

McMaster et al. (1975) reviewed a large series of hemangiopericytomas involving all regions and placed them into several histologic categories. Low grade (benign) hemangiopericytomas were characterized by a prominent vascular pattern and little vascular compression by spindle shaped tumour cells. They had rare mitotic figures without any haemorrhage or necrosis. All their patients with a low grade hemangiopericytoma were alive five years after treatment. Intermediate grade hemangiopericytomas were characterized by a less prominent vascular pattern and less spindle shaped tumour cells that caused more vascular compression. Mitoses were present but not abundant. In this class, the survival at five years fell to approximately 52%. High grade (malignant) hemangiopericytomas had a high degree of anaplasia with abundant mitoses. They also exhibited tightly compressed vascular spaces. Five year survival of this group was approximately 25%, with distant metastases seen in almost 65% of the cases (McMaster et al., 1975). However, none of these criteria have proven to be reliable in predicting the biological behaviour of hemangiopericytomas (Batsakis, 1979; Gudrun, 1979; Anderson, 1984; De Campora et al., 1985; Blitzer et al., 1985).

In general, hemangiopericytomas manifest a metastatic rate ranging from 12–60%. Interestingly, tumours that originate in the sinonasal tract demonstrate a lower metastatic rate of only 5–10% (Batsakis, 1979; Gudrun, 1979). Regional lymph node involvement is very unusual. The lungs and skeleton are the most frequent sites for distant metastases (Batsakis, 1979). The local recurrence rate for hemangiopericytomas of the nasal cavity ranges from 8% to 53% (Batsakis and Rice, 1981; Batsakis et al., 1983; Blitzer et al., 1985). Recurrences are thought to occur because of inadequate excision and may not appear until many years after treatment.

Various methods of treatment have been employed in the past. Most authors agree that the treatment of choice for hemangiopericytomas of the nasal cavity is surgical excision (Batsakis, 1979; Gudrun, 1979; De Campora et al., 1983; Gupta et al., 1985). Due to the high degree of vascularity, there is usually a substantial blood loss during biopsy and surgical removal of the tumour. It is advisable that biopsy of tumours suspected to be of vascular origin be done with caution and preferably in the operating room. Chemotherapeutic agents such as Adriamicin, alone or in combination have been used with only limited success in treating these tumours (Batsakis, 1979; Gudrun, 1979; De Campora et al., 1983). Wong and Yagodo (1978) report a 40% rate of partial or complete remission after treatment with this regimen. Radiotherapy has been used in several cases for palliation or as adjuvant treatment without demonstrating any improved survival compared to surgery alone. (Batsakis, 1979; Gudrun, 1979). Radiotherapy alone is reported to have a cure rate of only 13.3% (Batsakis, 1979).

The case presented here illustrates several characteristic features of hemangiopericytomas. The clinical presentation of a slowly growing, non painful, nasal

mass causing obstruction and an external nasal deformity is typical, although not unique to hemangiopericytomas. The diagnosis of hemangiopericytoma in this case could not be made on the basis of clinical appearance. The histologic diagnosis was not able to be made by routine Hematoxylin and Eosin staining, but required special reticulin stains. The diagnosis of hemangiopericytoma was confirmed by electron microscopy.

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