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Nasal Hemangiopericytoma

— A Case Report —

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A case of rare intranasal hemangiopericytoma in a 68-year-old male was reported with typical histological and ultrastructural findings. The lesion clinically resembled a nasal polyp. The experience in the present case raised the need for differential diagnoses of vascularised spindle cell tumors and of traditional hemangiopericytoma in soft tissue.

Key Words: *Hemangiopericytoma, Nasal cavity*

INTRODUCTION

Hemangiopericytomas are rare vascular tumors believed to be derived from the pericytes (Stout and Murray, 1942), and they usually occur in the retroperitoneum, the thigh, and the region of the head and neck. This type of tumor was first described by Stout and Murray in 1942 (Stout and Murray, 1942). In the traditional soft tissue form it has a well established microscopic pattern and a reasonably predictable behavior based on microscopic criteria (Enzinger and Smith, 1976). However, the diagnosis of this tumor and prediction of its clinical behavior still cause considerable problems.

Groups of hemangiopericytoma-like tumors of the nasal cavity were reported (Eneroth et al., 1970; Compagno and Hyams, 1976; Compagno, 1978; Gündrün, 1979; Batsakis and Rice, 1981; Batsakis et al., 1983; Tadwalkar et al., 1984; Chawla and Oswal, 1987; Eichhorn et al., 1990). These tumors were a peculiar form of vascular neoplasm probably related to traditional hemangiopericytomas, but with somewhat different morphologic features, location, clinical setting and biological behavior.

Recently, the authors encountered a case of nasal hemangiopericytoma. This article presents a detailed discussion of the histological features and the differential diagnosis using immunohistochemistry and electron microscopy.

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CASE HISTORY

A 68-year-old male was admitted to the Department of Otolaryngology with a history of profuse nasal bleeding and obstruction. Three years ago, a polypectomy was performed with the impression of the nasal polyp. The subject was in good health except for intermittent nasal bleeding. A paranasal sinus series revealed a polypoid lesion in the left nasal cavity. Mass excision was performed with the clinical impression of the nasal polyp.

PATHOLOGIC FINDINGS

The received specimen had fragmented pieces of gray-white solid tissue with fish-fleshy consistency and focal hemorrhage. The specimen measured 3 cc in volume. Microscopic examination revealed the tumor mass to be a circumscribed or pseudo-lobulated vascular neoplasrn characteristically composed of tightly packed cells surrounding the thin-walled endothelial lined spaces (Fig. 1). Many vascular channel of various sizes were embedded throughout the lesion. The surface of the tumor was often covered with either ciliated respiratory or benign metaplastic squamous epithelium. The individual tumor ceiis around the vascular spaces were uniform in appearance and elongated with indistinct cytoplasmic borders. In areas, spindle shaped cells were predominant and areas of myxoid change were present. Reticulin preparation revealed dense reticulin meshwork surrounding vascular channels and individual tumor cells (Fig. 2). Mi-

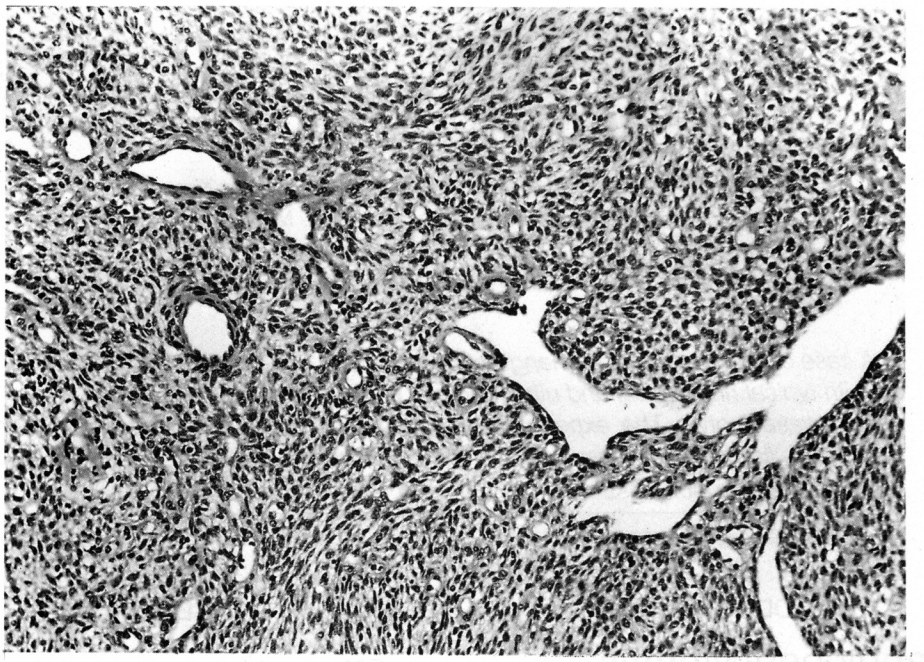

Fig. 1. Photomicrography of the tumor shows tightly packed cells surrounding the thin-walled endothelial lined spaces (H-E, ×100).

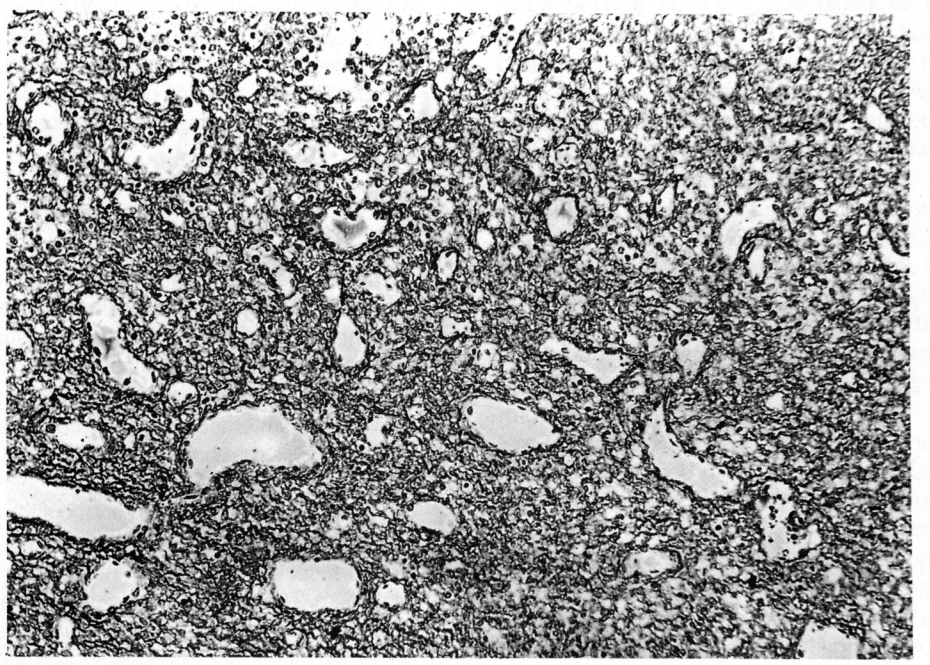

Fig. 2. Reticulin preparation reveals dense reticulin meshwork surrounding vascular channels and individual tumor cells (Reticulin, ×100).

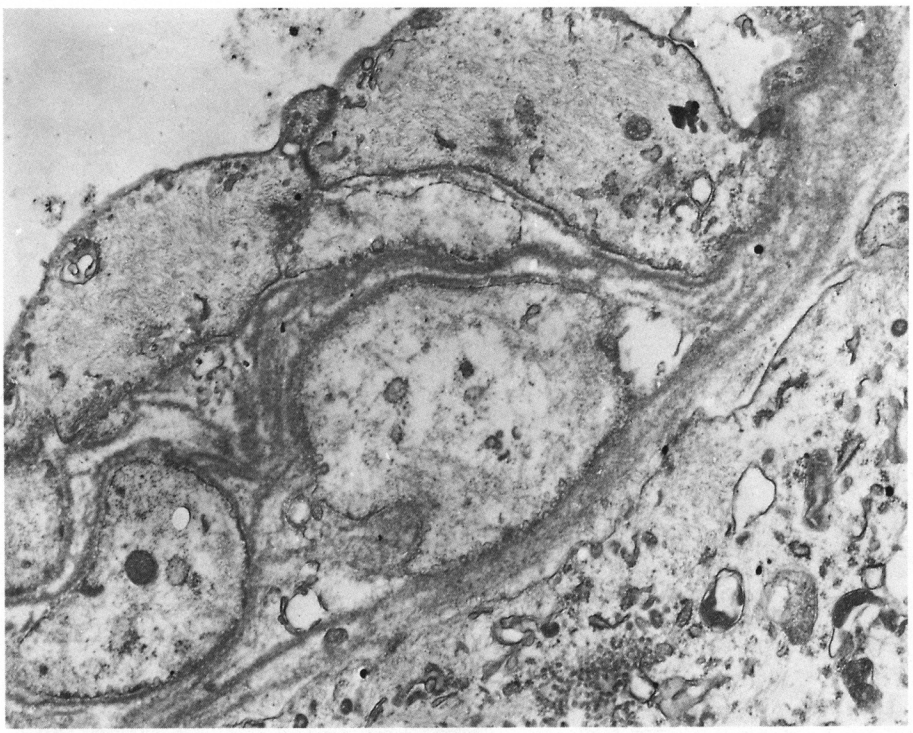

Fig. 3. Electron micrography of the tumor shows endothelial cells separated by duplicated or multilayered basal lamina (EM, ×10,000).

totic figures were rarely found, but some areas had a mitotic count of up to 2-3/10 high power field (HPF). Mast cells were noted throughout the tumor field.

Immunohistochemical staining of vimentin using the avidin biotin complex gave positive reaction to the tumor cells. However, S-100 protein and desmin revealed negative reaction.

Electron microscopic examination showed that the endothelium-lined vascular spaces were surrounded by the tumor cells. These spindle and stellate cells showed tapered cytoplasmic processes and oval nuclei. These cells were arranged in an orderly fashion around blood vessels. The tumor cells were segregated from the vascular endothelium by duplicated or multilayered basal lamina (Fig. 3). Organelles were found in few numbers and included mitochondria, Golgi apparatus, rough endoplasmic reticulum and free ribosomes and pinocytotic vesicles. Frequent microvillous extensions of the cytoplasm were found (Fig. 4).

DISCUSSION

Hemangiopericytoma is a rather uncommon neoplasm of soft tissue and arises from the proliferation of special cells called pericytes, which were described by Zimmermann (1923). These pericytes have been well established histologically as round or spindle-shaped cells with long branching processes; pericytes are applied to the outer wall of the venous capillaries and postcapillary venules. Since the first description by Stout and Murray (1942), hundreds of cases with similar microscopic features have been reported in various anatomic locations: the extremities, retroperitoneum, pelvic fossa, head and neck, and bone (Marcial-Rojas, 1960). In one series of 106 case analyses (Enzinger and Smith, 1976), the majority were found in the lower extremities and retroperitoneum. In McMaster et al. series (McMaster et al., 1975), the leading sites were the thigh and inguinal region. The first nasal hemangiopericytoma was reported by Stout in 1949 (Stout, 1949) in his second series of 25 cases. As pointed out in the literature (Eneroth et al., 1970; Compagno and Hyams, 1976; Compagno, 1978; Gündrün, 1979; Batsakis and Rice, 1981; Batsakis et al., 1983; Tadwalkar et al., 1984; Chawla and Oswal, 1987; Eichhhorn et al., 1990) occurrences of this tumor in the nasal cavity and paranasal sinuses are rare. Most of the tumors reported by Eichhorn et al. (1990) ap-

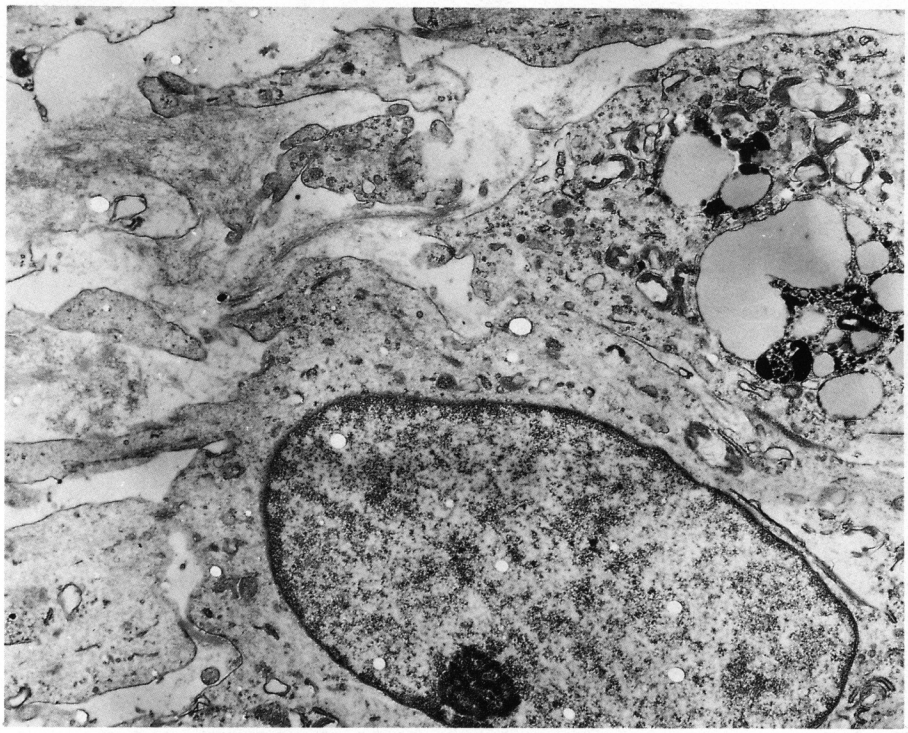

Fig. 4. The pericyte cytoplasm reveals sparse intracytoplasmic organelles and numerous cytoplasmic processes (EM, ×5,000).

peared to have arisen in the nasal passages; usually paranasal sinus was due to spread. This finding differs from the observation of Compagno and Hyams (1976) that paranasal sinus origin, with secondary usual involvement, was more common.

The correlation between histological appearance and grade of malignancy is usually poor (Eneroth et al., 1970). In some series, anatomic distribution had a bearing on malignancy, but this has not been completely substantiated (Batsakis and Rice, 1981). According to Enzinger and Smith (1976), the malignant behavior is usually suggested for the tumor with increased mitotic rates, greater degree of cellularity, immature and pleomorphic tumor cells and foci of necrosis and hemorrhage. McMaster et al. (1975) in a review of 60 cases from the files of the Mayo Clinic expected malginancy in cases having a slight degree of anaplasia and one mitotic figure/10 HPF or a moderate degree of cellular anaplasia and one mitotic figure/20 HPF. In the upper respiratory tract, the tumor appeared to deviate from the course followed by traditional patterns (Compagno and Hyams, 1976; Gorenstein et al., 1978; Gündrün, 1979; Chawla and Oswal, 1987). Compagno and Hyams (1976) described clinical, microscopic and gross features of 23 cases of intranasal hemangiopericytoma-like tumors and interpreted these lesions as a peculiar form of vascular neoplasm within the histologic spectrum of traditional hemangiopericytomas. These tumors were microscopically characterized by absence of mitotic activity, clear distinction of normal vessels from tumor cells, uniform spindle cells with little or no overlapping of cell borders, absence of necrosis and the presence of scattered mast cells. Follow up data showed no evidence of a malignant or biologically unpredictable lesion. However, Eneroth and associates (Eneroth et al., 1970) reported cases with local recurrence and late metastasis. It appears that the subject of this report is well concordant with the hemangiopericytoma-like intranasal tumor described by Compagno and Hyams (1976) in that the tumor is composed predominantly of spindle cells, lacks nuclear or cytoplasmic pleomorphism, has minimal mitotic activity, and has an absence of hemorrhage or necrosis. Even though a distinction between benign and malignant hemangiopericytoma is often difficult, the features described above led to good and favorable predictions of behavior in this case.

The immunohistochemical profile of this subject was similar to that reported by Winek et al. and Eichhorn et al. (Winek et al., 1989; Eichhorn et al., 1990). It also corresponded to a breast hemangiopericytoma that was immunostained (Mittal et al., 1986). In this case and from reports (Mittal et al., 1986; Winek et al., 1989; Eichhorn et al., 1990), tumor stained strongly only for vimentin; and it was negative for desmin and S-100 protein. However, in other reports (Winek et al., 1989; Eichhorn et al., 1990), S-100 protein was stained, unexpectedly.

Ultrastructural features of hemangiopericytomas from different anatomical locations have been reported (Nunnery et al., 1981; Ghadially, 1985). The most consistent features in Nunnery et al. series (1981) were the presence of basal lamina or basal lamina-like material that at least partly surrounded tumor cells and completely separated tumor cells from endothelial cells, tapered cell processes. These were found in our case.

Hemangiopericytoma may be confused with a number of tumors, (i.e. vascular leiomyoma, angiofibroma, glomus tumor, hemangioendothelioma, fibrous histiocytoma, mesenchymal chondrosarcoma and some endocrine tumors). In this case, angiofibroma, vascular leiomyoma and glomus tumor were the most important differential diagnoses. Angiofibromas is a locally destructive tumor composed of fibrovascular tissue of varying maturity, arising in or adjacent to the wall of the nasopharynx. The tumor occurs exclusively in young men and is much less cellular than the hemangiopericytoma-like tumor and contains a predominantly fibrous component. Immunohistochemical staining of S-100 protein and desmin was useful for the exclusion of leiomyoma and neurogenic tumor. Although immunohistochemistry and electron microscopy were useful, the diagnosis of hemangiopericytoma is entirely histological (Gündrün, 1979). Reticulin preparation revealed a dense reticulin meshwork surrounding the vessels and tumor cells similar to this case and this pattern was most helpful in delineating the spindle cell tumors.

In conclusion, the authors reported a case of nasal hemangiopericytoma and also found that hemangiopericytoma of the nasal cavity differed somewhat from traditional soft tissue hemangiopericytoma by its prominent spindle cell pattern, often with fascicular arrangement of the tumor cells, but with a lesser amount of interstitial collagen, and by a less intricate vascular pattern, histologically. The tumor is characterized by a relatively uncomplicated natural history and should be distinguished from true, traditional hemangiopericytomas. The tumor appeared to be effectively controlled with adequate wide local excision.

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