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Hemangiopericytoma in Sinonasal Region; A Case Report

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Abstract:

Hemangiopericytoma is a rare vascular tumor arising from skeletal muscle, although it can originate from any organ system. It is very rare in the sinonasal region. The main treatment is surgical excision. Clinical and radiological findings are not specific. In this case report, a patient with hemangiopericytoma in sinonasal region was described with current literature.

Keywords: ***Hemangiopericytoma, sinonasal region***

Introduction

Hemangiopericytoma is a rare mesenchymal neoplasm and accounts for less than 1% of all vascular tumors.[1,2] It was first described by Stout and Murray in 1942. The origin of the tumor is capillaries and pericardial cells located around the postcapillary venules, which regulate blood flow and permeability and were first described by Zimmermann.[3] The pericides are wrapped around all capillaries and are thought to be immature smooth muscle originating from the mesentery. Pericides play a role in blood flow regulation by providing structural support to capillaries. Although hemangiopericytomas can be found elsewhere in the body, they most commonly localize in the lower extremities, pelvic fossa, retroperitoneum, head and neck, chest and abdomen. We aimed to present this case with hemangiopericytoma in the sinonasal region with current literature.

Case:

A 57-year-old male patient was admitted to our clinic with a complaint of obstruction on the left side of the nose and an occasional nose bleed for about 6 months. Patient's rhinoscopy showed a polypoid tissue originating from the lateral wall of the left nasal cavity, which extended into the nasal cavity through the middle turbinate by penetrating it in the middle (Figure 1). There was no significant structure (such as a vascular network) noted. There were no other pathological findings in the otorhinolaryngologic examination. There was no obvious feature in the patient's medical history. Routine hematologic and biochemical parameters of the patient were normal. Paranasal sinus tomography scan of the patient revealed a lesion that filled the left nasal cavity, extending tocsuperior ethmoid cells and smooth-expanding and thinning lesions in the adjacent bones, extending to the frontoethmoid segment and continuing with soft tissue density in the left half of the frontal sinus (Figure 2, 3). Punch biopsy was taken from the lession with local anesthesia. Then, general anesthesia was performed to remove whole lession endoscopically.There was more bleeding than expected during the operation and bleeding was controlled with bipolar cautery during the operation. The postoperative pathology report was compatible with hemangioperistoma. No recurrence was observed in the follow-up period of 2 years.

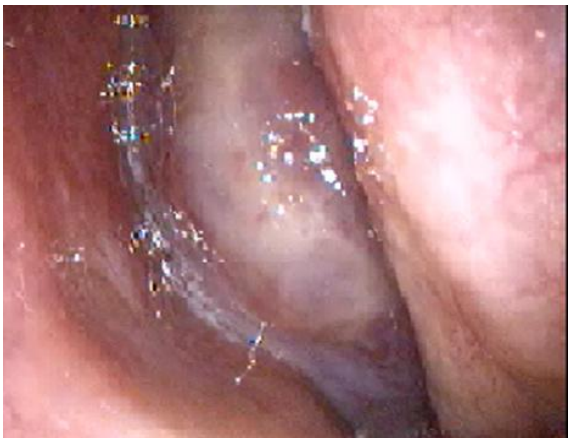

Figure 1: Mass seen in the left nasal cavity.

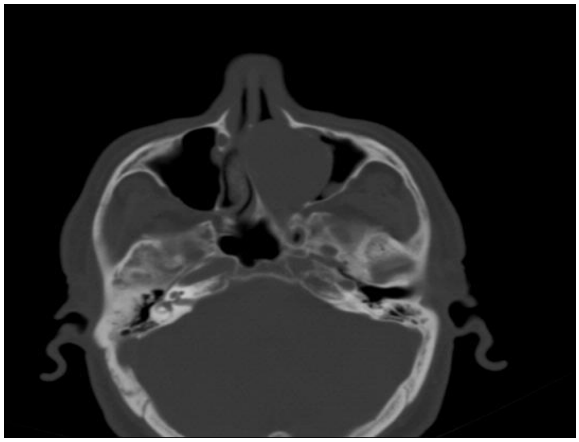

Figure 2: CT scan showing the mass in left nasal cavity

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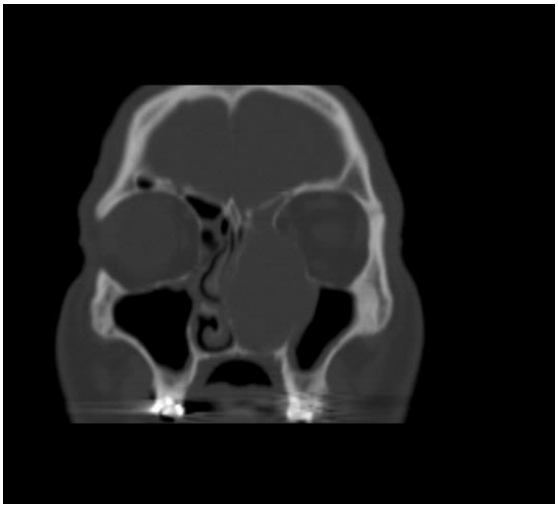

Figure 3: CT scan showing the mass in left nasal cavity

Discussion:

Hemangiopericytoma is a rare vascular tumor originating from pericapillary, contractile cells called Zimmermann pericides.[5] Trauma, prolonged use of steroids, hormonal causes have been suggested for etiology.[6] It is seen in head and neck region by 15-20%. The incidence of sinonasal region is less than 5%.[7] Hemangiopericytoma is most commonly observed in the abdomen and extremities. It can be seen in scalp, tongue, oral cavity, buccal mucosa, lip, nasal cavity and paranasal sinuses, maxilla, mandible, pharynx, parotid, orbita.[8] Sinonasal hemangiopericytoma is most commonly seen in the nasal cavity, ethmoid and maxillary sinus.

Nasal obstruction and epistaxis are the most common symptom of hemangiopericytoma. Pain, visual disturbances, headache are less frequent. Polyp of unilateral, edematous, hemorrhagic mass in red, pinkish gray, with bleeding can be seen in the examination. Bilateral involvement is rare.[9] In our case, nasal obstruction and epistaxis were complaints of our patient.

Hemangiopericytoma generally originates in the nasal cavity and may extend to the paranasal sinuses, although isolated paranasal sinus involvements have been reported.[10] After examination, imaging techniques must be carried out for differential diagnosis of mass lesions of the nasal cavity. Computed tomography scanning is always carried out and magnetic resonance imaging scanning may be necessary in some situations.[11]

Most hemangiopericytomas are benign. Enzinger and Smith have described malignant features in more than 4 large mitotic figures at 10 large enlargement sites as cellularity increase, nuclear atypia, hemorrhage, necrosis, and tumor size greater than 6.5 cm.[12] Benign hemangiopericytomas do not have mitosis, necrosis and hemorrhage. In addition to the obvious vascular structure, there are necrotic pericides.[12] Hemangiopericytoma can be seen in all age groups but increases in the 6th and 7th decades. Men and women are at equal frequency. Although benign, borderline, malign forms are described, hemangiopericytoma is accepted as a potential malignant.[13]

Patients usually have no pain in their clinic. Slowly growing painless mass is the most important symptom.[14] It may cause additional symptoms according to the location.[15] Various paraneoplastic syndromes such as hypoglycemia, hypophosphotemic osteomalacia, and hypertrophic pulmonary osteoarthropathy may also be associated with the mass.[16]

It is hard to seperate hemangiopericytoma from histiocytoma, chondrosarcoma, neuroblastoma, adenoid cystic carcinoma and mixed cell tumor, low grade fibromyxoid sarcoma (especially if the myxoid tissue is evident), juvenile hemangioma, glomus tumor, angiosarcoma, leiomyoma, leiomyasaroma, mesethelioma, synovial sarcoma.[17,18] Angiographic features may help differentiate hemangiopericytomas from other vascular lesions.[19]

The definitive treatment of hemangiopericytoma is surgical removal completely. Since the tumor does not have a real capsule, there is the risk of leaving a potential tumor cell behind. Up to now, there is no prospective study with radiotherapy or chemotherapy as the primary treatment. In the retrospective study of Backwinkel and Didams, in 15 patients treated with radiotherapy only 86.7% of the patients had recurrence within 5 years, but this rate was 46.9% in cases treated surgically.[20] The occurrence of advanced cases in the radiotherapy group also contributes to the inability to fully understand the role of radiotherapy in treatment. The use of radiotherapy is limited in cases with complete resection or irrespectable metastasis.[21] The results of studies on the role of chemotherapy in patients with metastatic disease are also uncertain.[22] Surgical resection was performed in our case and no recurrence was detected at 2 years follow-up.

As a result, the possibility of hemangiopericytoma should be considered in the polypoid and vascular originated masses in the sinonasal region and surgical resection should be performed with sufficient limits considering the high possibility of recurrence in case of detection.

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