CASE REPORT

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Malignant Schwannoma of Maxillary Sinus: A Case Report and Review of the Literature

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ABSTRACT Malignant schwannoma is rarely found in the paranasal sinuses and nasal cavity. Only 21 cases have been reported in the literature so far. Only four of the 21 cases were reported to be primarily originated from the maxillary sinus. In this article, a 44-year-old woman with a right maxillary sinus malignant schwannoma is presented. Right partial maxillectomy with lateral rhinotomy approach followed by a postoperative course of radiotherapy and chemotherapy was performed as the treatment. This mass was confirmed as a malignant schwannoma in histological examination and immunohistochemical staining. There was no tumor recurrence or distant metastases for 22 months after the treatment. She died from brain metastasis and sepsis 37 months after the operation.

Keywords: Malignant schwannoma; maxillary sinus

Malignant schwannomas are tumors that arise from the sheath of peripheral nerves, represent approximately l0% of all soft tissue sarcomas and are rarely found in the head and neck region.[1,2] Malignant schwannomas in the nasal cavity or paranasal sinuses are exceedingly rare and only 21 cases are reported at the paranasal sinuses and nasal cavity worldwide.[2,3] We present a case of malignant schwannomas of the maxillary sinus.

CASE REPORT

In August 2010, a 44 year old female patient with a history of nasal obstruction, rhinorrhea and epistaxis in the right nasal cavity for six months was presented. The patient complained of swelling on right side of face, right facial pain and exophthalmia in his right eye with unaffected visual acuity.

The right nasal cavity was full of hemorrhagic mass and nasal septum deviated to the left side on endoscopic examination. Right eye movements and vision were normal on the eye examination.

Computed tomography (CT) of the paranasal sinus revealed a mass in the right maxillary antrum ranging from the orbit, pterygopalatine fossa, nasal cavity and ethmoid cells. Magnetic resonance imaging (MRI) of paranasal sinus showed 75x40x50 mm sized gadolinium enhanced mass. There was no invasion of orbit and brain (Figure 1 a, b, c, d).

Right partial maxillectomy with lateral rhinotomy approach was performed with the Webermoore-Ferguson incision. The tumor did not infiltrate the orbital fat and muscle tissue. There was no tumor infiltration beyond the margin of resection. The right eye movements and vision were normal after surgery. Postoperative radiotherapy delivering 6996 cGy and adjuvant chemotherapy were administered to reduce the risk of the disease recurrence. Brain metastasis developed 22 months after the surgery. 2700 cGy by cyberknife radiotherapy was applied for the metasta-

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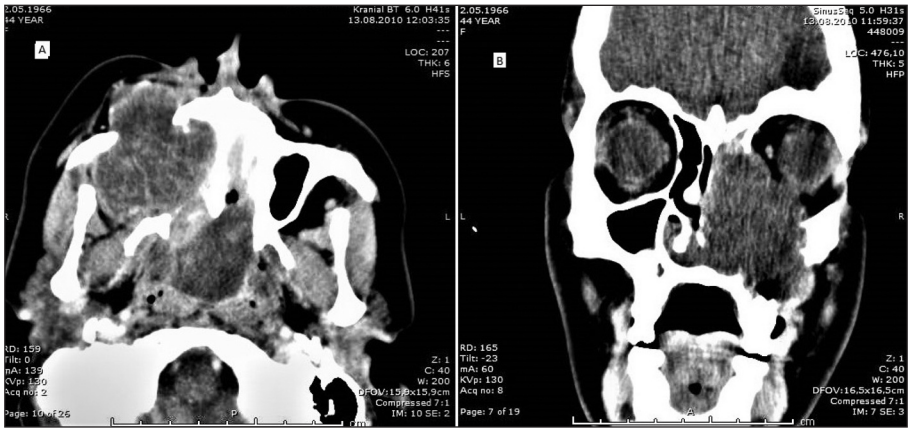

FIGURE 1 a, b: Computed tomography (CT) scan, paranasal sinus axial-coronal sections, there is a mass filling the right maxillary sinus, ethmoid sinus and nasal cavity. Computed tomography showing soft tissue mass causing medial and anterior wall destruction of the right maxilla.

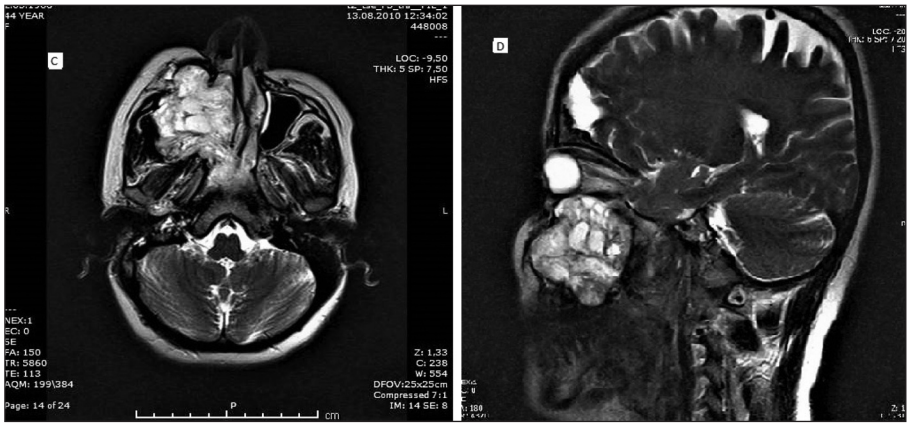

FIGURE 1 c, d: Axial and sagittal images of paranasal sinus T2-gadolinium enhanced magnetic resonance imaging showing 70x40x50 cm sized of right maxillary sinus. The tumor does not invade the orbita and the brain.

sis. The patient died from brain metastasis and sepsis 37 months after the operation.

Histopathological examination revealed a malignant schwannoma represented by atypical fusiform cells in fascicular arrangement with great alternation of cellularity H&E x40). Immunohistcochemical staining of the tumor shows strong immunoreactivity for S-100 (Figure 2 a, b).

DISCUSSION

Schwannomas are tumors arise from the schwann sheath of the peripheral nerves. Although 25-45% of all schwannomas have been reported to occur at the head and neck region, sinonasal schwannomas are uncommon tumors and represent 4% of head and neck schwannomas.[4,5] The origins of sinonasal schwannomas are postulated to arise from the maxillary and ophthalmic branches of the trigeminal nerve (fifth cranial nerve). Malignant schwannoma originating from the nasal cavity and paranasal sinuses are extremely rare.[6] Millard and Busser reported the first case of malignant schwannoma originating from maxillary antrum in a 42-year-old female patient.[7] Since then, 21 cases of malignant schwannoma of nasal cavity and paranasal sinuses have been added in the literature.[2] Many studies have presented that malignant schwannoma at sinonasal tract is rare. Only four of the 21 cases were reported to be primarily originated from the maxillary sinus.

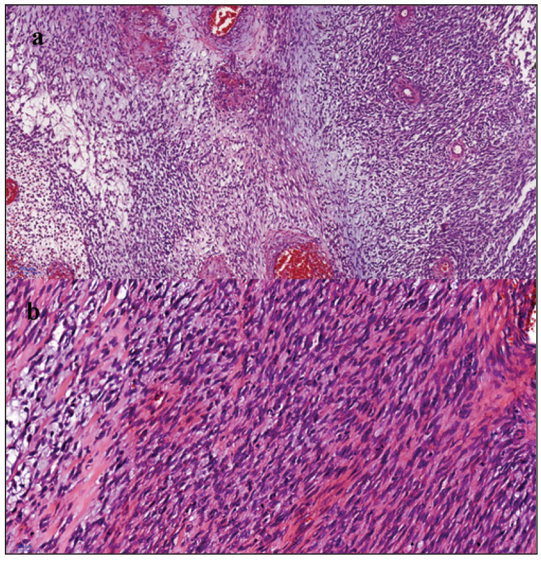

FIGURE 2a, b: Malignant schwannoma is a tumor consisting of cellular areas, tapestry appearance or cellular areas that change with a marble-like pattern. The tumor is cellular and has a fascicular growth pattern with mitosis. These folds are strongly S-100 positive.

Literature review revealed that nasal cavity and ethmoid sinus are more commonly involved than other sinuses.[5] Minhas et al. reported a case of malignant schwannoma originating from maxillary antrum in a 64-year-old male patient.[8] Khanna et al. found six cases of maxillary sinus schwannoma after literature search, one of which was malignant.[9] Only four of the 21 cases were reported to be primarily due to the maxillary sinus. In our case the tumor involved right maxillary sinus.

Malignant schwannoma can occur alone, but is associated with Von Recklinghausens disease in 30% of cases and usually evolves from malignant transformation of neurofibroma. Malignant schwannoma may arise as a result of malignant transformation of preexisting neurofibroma (Von Reckling hausen's disease). In our patient, there is no sign of preexisting neurofibromas. Hematogenous metastases at lung and bone have been reported in 33% of malignant schwannoma. Regional lymphatic metastases have rarely been reported. Elective neck dissection is not recommended because of the low probability of lymphatic spread.[1-6,7,10]

Imaging features of malignant schwannoma are generally nonspecific. Malignant schwannoma may show extensive infiltration. Bone destruction can cause and erosion; therefore, such a finding necessarily indicates malignancy.[2,3] In our case, erosion of bone was noted in computed tomography, a mass with hyperintense and increased vascularity was noted in MRI. Schwannoma is an encapsulated tumor with two histological variants described by Antoni A, shows hypercellularity (Antoni A, shows high malignancies), whereas antoni B has myxoid component. Schwannom exhibits intense immunostaining for S-100 protein (particularly in Antoni A areas) as usual. Histopathologically densely packed hyperchromatic spindle cells with slender wavy or curled nuclei with mitotic activity and indistinct cytoplasm suggestive of malignant schwannoma.[10] In our patient, S-100 protein showed positive reactivity.

Treatment modalities for malignant schwannoma consist of surgical excision, systemic chemotherapy and radiotherapy. Complete surgical excision with a safety margin is the mainstay treatment in patients with localized diseases. Radiotherapy and chemotherapy have been indicated in cases in which the tumor cannot be completely resected.[2,3,6] In our case, there was no lymphadenopathy in the neck and no distant metastasis was detected in the systemic scan. The main treatment for this tumor is en-block surgical resection as wide as possible.[1-7,9] We did not perform elective neck dissection. Postoperative radiotherapy and adjuvant chemotherapy were administered to reduce the risk of the disease recurrence. The five-year survival rate is 65.7% among individuals presenting malignant schwannoma alone, and falls by 30% when associated with type 1 neurofibromatosis.[1-3] Our patient died 37 months after the operation.

CONCLUSION

Malignant schwannoma in the paranasal sinuses or nasal cavity are extremely rare. Computed tomography and MRI of paranasal sinus help in diagnosis. The definitive diagnosis is only given through histological and immunohistochemical evaluations, with positive findings of S-100 protein. The treatment plan consists of gross total resection, radiotherapy and chemotherapy. Regardless of treatment, long time prognoses are very poor.

Written Consent

It was obtained from the patient who participated in this study.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Vasıf Soysal, Fatma Tokat;* ***Design:*** *Vasıf Soysal, Fatma Tokat;* ***Control/Supervision:*** *Vasıf Soysal, Fatma Tokat;* ***Data Collection and/or Processing:*** *Vasıf Soysal, Fatma Tokat;* ***Analysis and/or Interpretation:*** *Vasıf Soysal, Fatma Tokat;* ***Literature Review:*** *Vasıf Soysal, Fatma Tokat;* ***Writing the Article:*** *Vasıf Soysal, Fatma Tokat;* ***Critical Review:*** *Vasıf Soysal, Fatma Tokat;* ***References and Fundings:*** *Vasıf Soysal, Fatma Tokat;* ***Materials:*** *Vasıf Soysal, Fatma Tokat.*

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