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Malignant Peripheral Nerve Sheath Tumor of Frontal Sinus: A Case Report, Literature Review, and Immunohistochemical Study

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Abstract

Introduction: Malignant peripheral nerve sheath tumors are rare sarcomas derived from peripheral nerves or their sheaths. Head and neck region involvement in malignant peripheral nerve sheath tumors has been reported in only 15% of cases.

Case Presentation: A 45-year-old man presented to us because of left forehead area swelling. Computed tomography scan demonstrated a mass originating from left frontal sinus which was associated with bone destruction and soft tissue invasion. Post-operative histopathologic studies and immunohistochemical analysis of the lesion, confirmed «malignant peripheral nerve sheath tumor» as a diagnosis.

Conclusions: This case provides evidence that, although rare, malignant peripheral nerve sheath tumors can occur in frontal sinus. Combination of microscopic and immunohistochemical analysis is required for diagnosis of MPNSTs.

INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) are rare sarcomas [1], previously known as malignant schwannoma, neurofibrosarcoma or neurogenic sarcoma that the World Health Organization (WHO) replaced the term "MPNSTs" instead of previous terminologies [2]. The incidence rate of MPNSTs is 0.1/100,000 per year and they form almost 5% to 10% of all malignant soft tissue tumors [3]. MPNSTs have an association with type 1 neurofibromatosis, in which they are associated with this condition in 70% of patients, whereas the remaining of 30% occur spontaneously. MPNSTs are any malignant tumors arising from peripheral neurons or their sheaths [4]. Although radiation exposure is known as a risk factor, the etiology is unknown. MPNSTs generally happen between the third and fifth decades of life [1]. Although MPNSTs can occur in any anatomical region, the sciatic nerve is most common site which is affected. MPNSTs may involve face in rare cases [5]. Since MPNSTs can be derived from various types of cells, their presentation can vary from one person to another, and this makes the diagnosis difficult. Herein, we report a case of MPNSTs arising from frontal sinus in a 45-year-old man. To the best of our knowledge, only a few cases of MPNSTs arising from frontal sinus have been reported.

CASE PRESENTATIONS

A 45-year-old man presented to us because of left forehead area swelling, which had been begun from a few month ago. The patient had a history of a mass surgical excision (in front of the left ear), with sarcoma pathology one year ago followed by radiotherapy. The patient also complained of an occasional pulsating pain in the swelling site. The patient didn't have epistaxis, rhinorrhea or malodor. Systemic symptoms such as fever, weight loss or anorexia was not noted.

Upon physical examination, a mass, about 2 × 2 cm in size, without erythema or discharge existed in the left forehead area. It had no effect on the patient's vision. Neurological examination including second, third and seventh nerves was normal. In palpation, the mass was firm, fixed and without tenderness. The tympanic membranes were normal. Nasal septal deviation was not noted. The patient did not have any family history of neurofibromatosis.

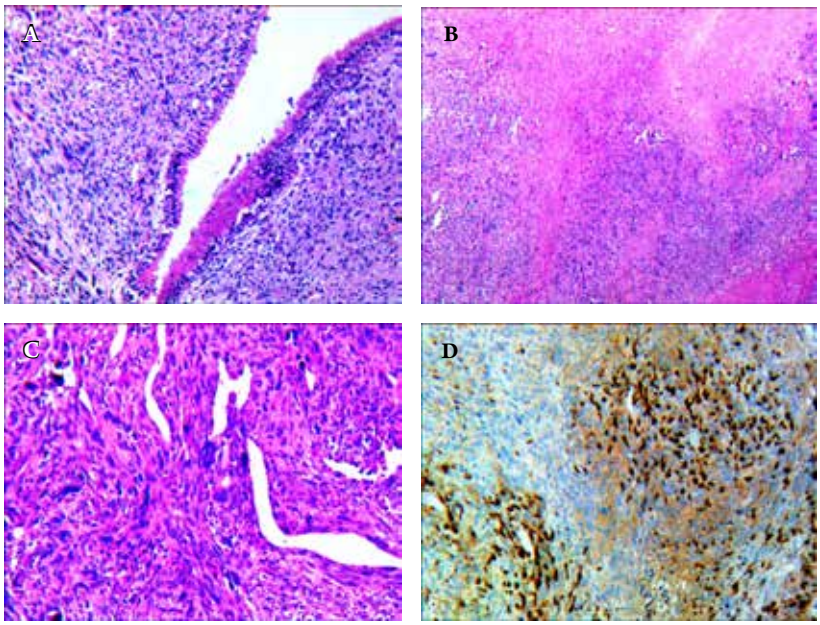

Figure 1: a). Shows extension of highly cellular tumor beneath the respiratory epithelium (H&E stain, x10); b). Geographic tumor necrosis is present. (H&E stain, x4); c). Reveals Hemangiopericytoma-like vessels. (H&E stain, x10); d). Immunostaining shows the tumor cells expressed focally S-100 protein. (IHC stain, x100).

Computed tomography (CT) scan without contrast, showed a mass with a diameter of 35 mm originating from left frontal sinus that was associated with bone destruction and soft tissue invasion. An opacity was visible in the left frontal sinus. Moreover, destruction of the posterior wall of left maxillary sinus was listed. In addition, mucosal thickening in both ethmoidal sinus was seen.

Magnetic resonance imaging (MRI) of sinuses with and without contrast revealed an enhancing lobulated mass, 22 × 12 mm in size, in the left frontal sinus and the adjacent bone destruction. Left anterior fossa invasion was observed, too. Fine needle aspiration cytology (FNAC) was positive for malignancy.

Under general anesthesia, a bicoronal incision was created by the surgeon and with anterior cranial fossa access, the mass had involved supra-orbital and supra-trochlear nerves. The mass involvement was evident to the posterior wall of frontal sinus, roof of the orbit and Dura matter. To ensure, middle frontal sinus wall was also removed and the ethmoid sinus mucosa sample was sent to frozen section that was negative. The Dura involving mass was resected and CSF leak was repaired and the anterior wall of frontal sinus was renovated with titanium mesh and transferred fat from the belly.

Resected specimen with margins was sent to pathology laboratory. Gross examination showed two pieces of cream-colored tissue measuring 4 × 3 × 2 cm. Sections revealed a tumor composed of oval to spindle cells, some with bent nuclei and arranged in densely packed whorled fasciculi. Cellular pleomorphism and mitotic activity were seen. There was extension of tumoral cells beneath the frontal sinus and respiratory epithelium. Other morphologic changes were hemangiopericytoma-like vascular proliferation and geographic necrosis (Fig. 1). Immunohistochemistry (IHC) findings in our case are summarized below (Table.1). The patient was followed up for 6 months without signs of recurrence or other complications.

Table 1: IHC Findings in Our Case

Marker	Status
Vimentin	Diffusely positive
S-100	Focally positive
GFAP	Positive in moderate number of tumoral cells
CD34	Positive
Desmin	Negative
SMA	Negative
CKAE1/3	Negative
CD68	Scattered positive cells

Abbreviations: SMA: Smooth muscle actin; CKAE1/3: Cytokeratin AE1 / AE3

DISCUSSION

MPNSTs are tumors typically originating from Schwann cells and sometimes from a peripheral nerve. MPNSTs may also develop from transformation of some lesions such as plexiform fibroma [6]. MPNSTs affect men and women with the same gender distribution [7]. The most common anatomical location involved is the trunk and limbs. In fewer cases, head and neck may be involved [1]. These tumors generally present as an enlarging palpable mass [8]. In our patient, MPNST was confined to the left frontal sinus area. Among MPNST's risk factors, neurofibromatosis type 1 and a history of radiation can be mentioned. MPNST may be associated with neurofibromatosis type 1 or it may be sporadic [9]. In this case, the patient didn't have neurofibromatoses in his body but with history of radiation during last year. Due to the morphological similarity of MPNSTs with other sarcomas and on the other hand, due to lack of molecular and immunohistochemical markers, the diagnosis is difficult [7]. A diagnostic criteria which is used is "when a sarcoma is named

Table 2: Some Previous Cases of MPNSTs Involving Head and Neck Region Which Are Described in Literature

Age/sex	Clinical presentation	Location	Treatment	Reference
74-year-old/ Male	Forehead swelling and right eye deviation	Frontal Sinus, orbital cavity and ethmoid sinus	Surgical excision of the lesion	[1]
55-year-old/ Female	Dizziness, vertigo, periorbital, frontal, and left cheek soreness, worsening diplopia, and weight loss	Left frontal sinus extending into the left anterior ethmoid air cells	Wide-local excision and adjuvant radiotherapy	[8]
68-year-old/ Female	Visual problems such as exophthalmos and blurred vision	Orbit	Total resection of tumor	[5]
24-year-old/ Male	Increasing pain, discharge from nostril and gradual weight loss	Maxillary sinus	Palliative radiotherapy and chemotherapy	[13]
12-year old/ Male	Pain in the superior maxilla	Maxilla	Hemimaxillectomy and radiotherapy	[14]
38-year-old/ Male	Pain in cheek, bleeding nostril, and nasal blockage	Maxilla	Total maxillectomy and planning of Adjuvant radiotherapy	[15]

MPNST that at least has one of the three following criteria: 1) the tumor is derived from peripheral nerve 2) the tumor arises from a benign pre-existing nerve sheath tumor which is usually a neurofibroma 3) the tumor shows Schwann cell differentiation in histologic studies" [10]. Frontal, Maxilla and ethmoid sinuses as well as, orbit involvement has been reported previously (Table 2). To the best of our knowledge, only few cases of MPNSTs involving frontal sinus have been reported. Surgery is used to treat MPNSTs of the head and neck [8]. Previous studies have noticed the role of radiotherapy as adjunctive treatment [11, 12]. In our case, the patient underwent adjuvant radiotherapy after surgical resection. The patient was followed up for six months without signs of recurrence or other complications.

Malignant peripheral nerve sheath tumors are rare tumors of neurogenic origin. Although MPNSTs are rare sarcomas, but clinically important condition which proper management may improve prognosis. MPNSTs can involve frontal sinus in rare cases. Combination of microscopic and immunohistochemical analysis is required for diagnosis of MPNSTs.

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CONFLICTS OF INTEREST

Authors declare that there is no conflict of interest.

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AUTHORS' CONTRIBUTIONS

Mojaba Babaei Zarch involved with writing the first draft of article and submission. Mansour Moghimi involved with reporting pathology and writing the article. Mohammad hossein Dadgarnia involved with performing surgery. Seyed Mojtaba Ghelmani involved with writing article and revising. Mohammad Baghi Yazdi and Fatemeh Imani involved with correcting of English mistakes and add table.

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