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Sinonasal teratocarcinosarcoma

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Abstract

Teratocarcinosarcoma is a rare, morphologically heterogeneous and highly malignant neoplasm. It is characterized by the presence of benign and malignant epithelial, mesenchymal and neural components. The carcinoma may be either squamous or adenocarcinoma and the mesenchymal component may manifest spindle, smooth, skeletal muscle, cartilage and bone features. Because of their infrequency, these lesions are often misdiagnosed, leading to management difficulties. In this case report we have shared our experience with sinonasal teratocarcinosarcoma in a 23 year old female and performed a brief review of literature.

Keywords: Sinonasal teratocarcinosarcoma, Pakistan, Malignant.

Introduction

Sinonasal teratocarcinosarcoma (STCS) is an extremely rare malignant tumour, thought to arise exclusively in the sinonasal tract. It was first described by Shanmugaratnam in 1983 as teratoid carcinosarcoma, consisting of various components of neuroectodermal, epithelial and mesenchymal proliferations of varying maturity. Keeping its complex cyto-architecture in mind the tumour was later named as sinonasal teratocarcinosarcoma by Heffner et al in 1984, after their review of 20 cases. The tissue heterogeneity usually leads to misdiagnosis especially where limited sampling is carried out. Of the 20 cases reviewed by Heffner, many were mislabeled as adenocarcinoma, olfactory neuroblastoma, fibrosarcoma and rhabdomyosarcoma.

A review of literature by the authors found 88 cases of this tumour, as case reports and short series, associated with loco-regional recurrence and high mortality. One of the larger series calculated a two year disease free survival rate of 28% and the overall survival rate of 46%. However, most of the literature deals with histopathological findings and tissue architecture of the tumour leaving a void with respect to clinical findings, management and treatment outcomes. Due to the rarity of this condition management guidelines are currently unavailable and management is tailored to suit the patient. Here we present our first experience sinonasal teratocarcinosarcoma in a 23 year old female and a corresponding review of literature.

Case Report

A 23 year old female presented to our Otorhinolaryngology clinic with complaints of a mass protruding from her right nostril. She was in her usual state of health four months ago when she started experiencing nasal obstruction and epistaxis. Within a matter of days, she developed a mass protruding out of her right nostril, which was partially excised at another center intranasally. Histopathological examination revealed STCS (Figure-1). Fifteen days after its excision the mass reappeared.

On examination a reddish growth was seen protruding from her right nasal cavity, completely obstructing the right nostril. The mass was soft, friable, and fleshy in appearance. On the external aspect the mass was approximately 1.5 x 1.5 cm; however clinical assessment of its size could not be conducted as the bulk of the tumour was within the nasal cavity and paranasal sinuses. Magnetic resonance imaging (MRI) with contrast showed a large exophytic mass involving the right nasal cavity. The mass was bulging posteriorly almost obliterating the nasopharyngeal space. Superiorly it was involving the ethmoid air cells and was

Figure-1: Teratocarcinosarcoma of the nasal cavity showing an admixture of well-formed glands with atypical epithelium along with sarcomatous areas and primitive neuroepithelium with rosettes. (H&E, 4X).
closely adherent to dura; however no dural breach was noted. Significant obstruction of the right maxillary sinus and sphenoid sinus bilaterally leading to mucosal thickening and fluid retention was also seen (Figure-2). There was no cervical lymphadenopathy.

A right lateral rhinotomy and maxillectomy was performed and the tumour was excised. An attempt was made to remove mass in a single piece. However, near the dura it was removed in a piece meal fashion with the help of an operating microscope (Carl ziemss; Neurosurgical adjustable microscope, upto 400 magnification). This careful piece meal removal was done to avoid disruption of the dura. Bleeding was moderate during surgery and multiple bleeders were identified which were controlled using bipolar cautery. Near dura adrenaline soaked patties were also used along with bipolar cautery. Estimated intraoperative bleeding was around 200 ml. The mass was adherent to nasal septum, maxillary and ethmoid sinus walls making it difficult to ascertain the site of origin. During the excision the patient developed a dural tear near the cribriform plate, which was repaired by our neurosurgical service by intranasal route. Post operative bilateral anterior nasal packing with bismuth iodine paraffin paste was done and a lumbar drain was also placed. Her postoperative recovery was unremarkable with no need of blood transfusions. The nasal packs were removed after 48 hours and the lumbar drain was removed on the 3rd postoperative day following which she was discharged.

She was planned for concurrent chemo-radiotherapy and six cycles of Cisplatin (20 mg/m² days) and Etoposide(100 mg /m² days) with 35 fractions of radiation (external beam radiation using bi-linear accelerator, upto 70 Gy) were given. An excellent clinical response was achieved. The patient remained under regular follow up for last 6 months, with no recurrence.

Discussion

Sinonasal teratocarcinosarcoma is thought to arise from the pluripotent olfactory epithelial cells in the nasal cavity. However, some ambiguity exists regarding the cells of origin. Most authors agree that a pluripotential progenitor cell with multidirectional differentiation capabilities is the most likely case giving rise to the complex cyto-architecture and tissue heterogeneity. The tumour is composed of neural, epithelial and mesenchymal neoplastic tissue in varying degrees of maturity. Due to the presence of multiple cell lines STCS may mimic other pathologies derived from those cells, depending upon the predominant tissue obtained at biopsy. Limited biopsy samples may therefore fail to establish any diagnosis. The presence of foetal squamous cells which exhibit clear cytoplasm is however, considered a pathognomic histological feature of the disease.

These highly malignant tumours initially present with relatively benign complaints of recurrent epistaxis (53.52% of cases) and nasal obstruction (61.97% of cases). Other manifestations raising suspicions of malignancy, such as odynophagia, dysphagia, expectoration of tissue, epiphora, headache, vision loss, exophthalmos, anosmia and altered mental status arise when the tumour invades surrounding tissues and the severity if related to the degree of tumour extension. Cases of patients presenting with syndrome of inappropriate anti diuretic hormone secretion (SIADH) have also been reported. In our analysis of available literature these symptoms presented within a short mean duration of 102 days, which could be attributed to the aggressive nature of the tumour.

On nasal examination a moderately firm red or reddish-purple mass may be visualized with friable and necrotic areas; manipulation of the mass may lead to epistaxis. The findings were consistent with our case with a red-coloured mass protruding from the nasal cavity which bled on examination/manipulation. Most common sites involved are the nasal cavity and paranasal sinuses. Bone erosion and intracranial extension were also noted in 24.19% and 20.96% of the reported cases at the time of presentation respectively.

Analysis of the available literature on STCS showed a strong male predominance with an average age of presentation at about 51.75 years. However of interest is that our patient was the third reported patient under the age of 25, while the other two cases arose from India, indicating that a common environmental or genetic link for an early age of onset of this disease.

Currently no management guidelines are available.
regarding the disease and most of the literature focuses on the histo-pathological findings. Clinical details were available for about half of the reported cases, analysis of which revealed that 87.14% of cases underwent aggressive surgical excision followed by adjuvant radiotherapy in 60.00%. About 11.42% of patients also underwent adjuvant chemo/radiotherapy post-surgically. However local recurrence after excision is high with reported 3 and 5 year survival rates at about 30% and 20%, respectively. In our analysis of literature we found recurrence in 42.68% of the cases with a mean recurrence time of 21.3 months in patients followed for a mean period of 76.1 months.

The high rate of recurrence is suggestive of the aggressive biological nature of the disease and although the prognosis does not appear encouraging aggressive surgery followed by chemo-radiation appears to be the mainstay of management for this condition.

References