ruled out. These can clinically present as swellings with no associated symptoms which can be misdiagnosed as a neoplastic process. Most of the cases reported in the literature have been wrongly misdiagnosed as tumors, but FNAC has resolved the issue in every case^{1, 2}. These patients have been found to have no recurrence on a follow up. The present case also received symptomatic treatment and was found to be free of disease after a follow up of 15 months.

An awareness of such an occurrence is important for both cytopathologist and the clinician. FNAC plays a major role in the diagnosis of these lesions and avoids unnecessary surgeries.

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SOLITARY MALIGNANT SCHWANOMA OF PARA PHARYNGEAL SPACE -A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT : *Malignant Schwannoma is an aggressive soft tissue tumor that is more commonly found in association with Von Recklinghausens disease. The solitary tumors although rarer have a better prognosis when compared to those associated with Von Recklinghausens disease. Parapharyngeal tumors are rare and a majority of these are benign salivary or neurogenic tumors. A malignant Schwannoma at this site is very infrequent with only four cases reported so far. The case of a 16 year old girl with a parapharyngeal malignant schwannoma is presented for its rarity and our experience in dealing with it. A combined modality of treatment with surgery, radiotherapy and chemotherapy was used. Pertinent literature is reviewed.*

Key Words : *Malignant Schwannoma, Parapharyngeal Space Tumors*

INTRODUCTION

Parapharyngeal tumors constitute 0.5% of all head and neck tumors [Kerry Olsen] and although 20-40% of these are of neurogenous origin they are mostly benign schwannomas. About 2% of all neurogenous tumors turn out to be malignant. Malignant schwannomas (MS) arise in patients with and without Von Recklinghausens (VR)

Disease. The risk of developing malignant neurogenous tumors is 4600 times more in patients with Rr disease than in the general population [Ducatman et. Al].

In a retrospective study done at the Mayo Clinic, 120 cases of MS were detected over a period of 71 years. Of these 23 arose in the region of the head and neck (an

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Fig. I : Picture showing a diffuse swelling over the region of the cheek and submandibular area.

Fig. II : Digital subtraction angiographic picture showing tumor displacing carotid vessels posteriorly.

Fig. III : Operative photograph showing extent of the tumor.

annual incidence of 1.7%) [Ducatman et. al]. The present report is the first case of a Malignant Schwannoma that we have come across over the last 15 years during which we have dealt with 27 parapharyngeal tumors.

Fig. IV : Histopathological picture showing a cellular tumor with spindle to oval cells in fascicles with a mitotic figure.

Fig. V : Post operative picture of the patient.

CASE REPORT

A 16 year old girl presented at the out-patient department with complaints of dysphagia, mass in oral cavity and a swelling of the left side of the face of 6 months duration. A history of decreased hearing in the left ear as well as snoring at night was elicited. She had undergone tonsillectomy elsewhere two months prior to presentation to us. There was no family history of VR disease.

On examination there was a diffuse swelling over the region of the left cheek and submandibular area that was tender to touch, firm in consistency, not pulsatile and not compressible [Fig. I]. The examination of the oral cavity was limited by trismus. The left tonsillar fossa and the lateral wall of the pharynx was pushed medially. She had no detectable neurofibromas or café'au lait spots on her body. There were no neurological deficits and all cranial nerves were normal.

Routine biochemical and hematological investigations as well as urinary V.M.A. were within normal limits. Computed tomography confirmed a nonvascular tumor involving the left parapharyngeal space and extending on to the left infratemporal fossa. Digital subtraction angiography revealed the tumor to be pushing the carotid vessels posteriorly and receiving blood supply from the facial and lingual arteries [Fig. II].

The patient was taken up for surgical excision by a surgical team headed by the senior author, under general anaesthesia. The Modified Billers approach with a paramedian stepped mandibulotomy was employed to gain access to the tumor mass. The tumor was found extending laterally into the infratemporal fossa and superiorly up to the skull base, eroding the pterygoid plates and infiltrating the pterygoid muscles and the hard palate (Fig III). The nerve of origin could not be identified. Wide excision of the tumor was completed. Histopathology of the biopsied specimen revealed a cellular tumor with spindle to oval cells in fascicles, and at places palisading, interspersed with areas of necrosis with 2-3 mitoses per high power field (Fig. IV). As the patient developed left sided paralysis of the tongue postoperatively, it was concluded that the tumor arose from the left hypoglossal nerve.

Four weeks postoperatively she was subjected to a course of radiotherapy. She was given a total dose of 60 Gys of radiation in 30 fractions over a period of 6 weeks. This was followed by 6 courses of chemotherapy with Adriamycin, Cyclophosphamide and Vincristine at 3 week intervals. After completion of this cycle, a second cycle of 12 courses of vincristine, actinomycin-D and cyclophosphamide was completed. The patient was asymptomatic for 20 weeks after surgery but was readmitted for management of pleural effusion that she had developed during the course of chemotherapy. This was treated by pleural tapping and pleurodesis with Bleomycin. She is now under regular follow-up for 3½ years after diagnosis without any evidence of recurrence of the primary tumor or distant metastasis [Fig. V].

DISCUSSION

Malignant Schwannomas have been known to occur in almost any part of the body involving nerves of the extremities, abdomen, pelvis, chest, head and neck. In studies involving a large number of cases, the incidence of this tumor in the head and neck varies between 8-14% [Baillet et. al]. Only 4 cases of malignant Schwannomas

of the parapharyngeal space have been reported in the english literature over the last 20 years. This is not surprising considering the rarity of this pathology in this anatomical location.

The diagnosis of a neurosarcoma is rarely possible prior to surgery. The recognition and classification of a malignant sarcoma as being of nerve sheath origin is often difficult as the tumor may either grow outward from the involved nerve or may be intertwined with the nerve involved [Wilson et al]. Differentiating this tumor from other types of spindle cell sarcomas such as fibrosarcomas, leiomyosarcomas etc., is difficult with the routine techniques of histological analysis. D'Agostino et. al, after reviewing 104 cases of MS failed to reach a conclusion on the characteristic histopathology. It has been emphasized that the most important feature distinguishing a neurogenic sarcoma from a fibrosarcoma is its origin from a nerve trunk and this is not always easy especially in a malignant Growth. The malignant nature of the tumor is confirmed histologically by a spindle cell tumor showing histological areas of a neurofibroma, the presence of more than a rare or isolated mitosis and epineural invasion from within as seen under the electron microscope.

Although patients with VR disease are more prone to have MS as compared to the general population, the large series studies show MS in patients without VR disease to be more common [Kyriakos.M]. Stout was the first to link MS with VR disease and he made an intelligent guess of 5% of cases of VR disease may develop MS [Hutcherson et. al]. However different studies have given incidence varying from 3-30%. Women have been more commonly affected in those few reports that mention neurosarcomas in the head and neck but with the small number of cases reported its significance is doubtful.

Parapharyngeal tumors have to reach a size of 2.5-3. cms., before they can be clinically detected [Kerry Olson]. The malignant tumors of this region are manifested by symptoms due to the infiltration of the adjacent neurovascular structures. This may present as pain, neurological deficits, neuralgias, bradycardia, hypertension and trismus depending on the anatomical structures involved. The large size of the tumor may cause pharyngeal obstruction and eustachian tubal dysfunction with all the attendant consequences.

Complete surgical excision is the primary modality of

treatment for malignant schwannomas. It is often not possible to obtain clear surgical margins in the head and neck because of these tumors lacking circumscription and the anatomical constraints of this region. This is a possible causative factor for the lower survival rates in these patients. The modified Billers approach adopted by us, is unequaled in the quality of access and exposure obtained in the case of large tumors of the parapharyngeal space and the skull base.

Historically MS was considered to be radioresistent. In a study of 232 patients, Das Gupta found that most of his patients who had undergone primary radiation therapy required surgical treatment later. The limited response obtained by him may reflect older techniques rather than tumor behavior. Radiotherapy now, in almost all cases, has at least a palliative response and is definitely indicated for post-operative control of local disease. The effectiveness of chemotherapy in controlling the disease is still being studied. Sordillo et. al reported 100% 3 year survival in three of their patients treated with surgery and chemotherapy and 9 patients who received adjuvant chemotherapy after surgery for a primary, metastatic or recurrent disease were alive 3½ years after diagnosis. This is an encouraging development which offers scope for further research. Our patient who received post operative radio and chemotherapy is alive 3½ years after diagnosis, inspite of the large size and location of the tumor in the neck.

The prognosis in these cases is adversely affected by factors like, the presence of VR disease, poorer differentiation, higher rates of metastasis, multifocality, size larger than 5 cms. and the extent of tumor resection permitted by the anatomical constraints of the region. The patients who have a smaller tumor and undergo total excision have a statistically significant improved survival [Ducatman et. al].

CONCLUSION

Our case is one of the very few reported solitary malignant schwannomas of the parapharyngeal space. A combined modality of treatment with primary wide surgical excision followed by radiation and chemotherapy is a possible route to success in the management of these aggressive tumors.

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