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Case Report

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Undifferentiated carcinoma of parapharyngeal space in 8 year old female child: a rare case presentation

Ramesh Kadela[1], Sonia Jindal[1], Pooja Arya[1], Niranjan Nagaraj[2]*, Deepchand Lal[1]

[1]Department of ENT, S P Medical College, Bikaner, Rajasthan, India
[2]Department of Paediatrics, S P Medical College, Bikaner, Rajasthan, India

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***Correspondence:**
Dr. Niranjan Nagaraj,
E-mail: getniranjan806@yahoo.com

ABSTRACT

Parapharyngeal space tumors represent only 0.5% of all head and neck tumors. Out of which, 80% are benign and 20% are malignant. Tumor must be at least 2 cm in size before bulge or abnormality is palpable. We report a case of 8 year old female child who presented with bulge in right tonsillar region.

Keywords: Undifferentiated carcinoma, Parapharyngeal space, Pterygoid space

INTRODUCTION

Tumors of parapharyngeal space are rare, accounting for 0.5% of all head and neck tumors, of which approximately 80% are benign.[1,2] The salivary gland neoplasm represent the commonest tumour, followed by neurogenic tumour and paragangliomas.[3] Miscellaneous tumours are adenoid cystic carcinoma, mucoepidermoid carcinoma, squamous cell carcinoma, haem-angiopericytoma, fibrosarcoma, lymphoma, undifferentiated carcinoma and juvenile angiofbroma. Tumour size must be at least 2 cm in size to be detected clinically. Clinically, it presents as neck mass in 53% cases and oropharyngeal bulge in 51% cases. Imaging is done by CT, MRI and angiography. Main treatment modality is surgical excision. Radiotherapy and chemotherapy are used mainly for poor surgical candidates and unrepeatability. We provide an overview of parapharyngeal space tumours and present a case of undifferentiated carcinoma of parapharyngeal space.

CASE REPORT

An 8 year old female child presented to our OPD with complaints of difficulty in swallowing, difficulty in speech and swelling in right parotid region for 2 months.

Figure 1: Swelling in parotid region.

Figure 2: Bulge in right tonsillar region.

On clinical evaluation, swelling of approximately 3x3 cm in size, which is hard, non-tender, fixed to underlying structures is palpable in right parotid region (Figure 1). Intraorally, bulge is seen in right tonsillar region (Figure 2).

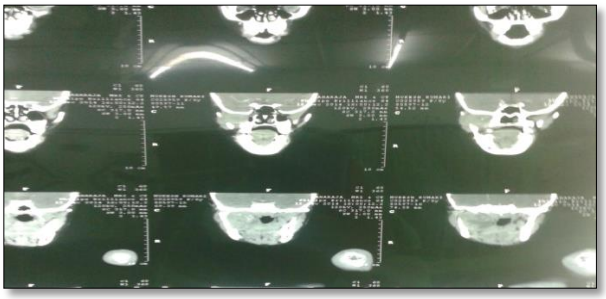

Figure 3: Coronal scan showing parapharyngeal tumour.

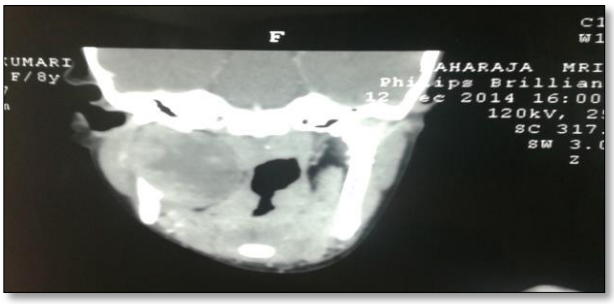

Figure 4: Lesion of size approximately 48x39x39 mm seen in right temporalis and pterygoid fossa.

Computed tomography scan was performed using submillimeter thin contiguous plain and contrast axial scan of neck from base of skull to clavicle. Coronal and sagittal reformatted images were obtained (Figure 3). Heterogeneously enhancing large well defined soft tissue density mass lesion of size approximately 48x39x39 mm seen in right temporalis and pterygoid fossa, with poor fat planes infiltrating into adjacent masticator muscles, erosion of condyle of mandible, extending up to medullary cavity, however, outer cortex are spared (Figure 4). Erosion of lateral and medial pterygoids and right postero-lateral wall of maxillary sinus also noted (Figure 5).

Figure 5: Erosion of lateral and medial pterygoids and right postero-lateral wall of maxillary sinus.

Bilateral mild tonsillar hyperplasia (R>L) noted. Few submandibular, cervical and supraclavicular lymphadenopathy noted, largest measuring approximately 15 mm.

An USG guided fine needle aspiration was done. On microscopic examination, smears show large cells in clumps. Individually cells are having scanty cytoplasm and round to oval hyper chromatic nucleus (Figure 6a and b). Occasional multinucleated giant cells are seen. These features are suggestive of undifferentiated carcinoma.

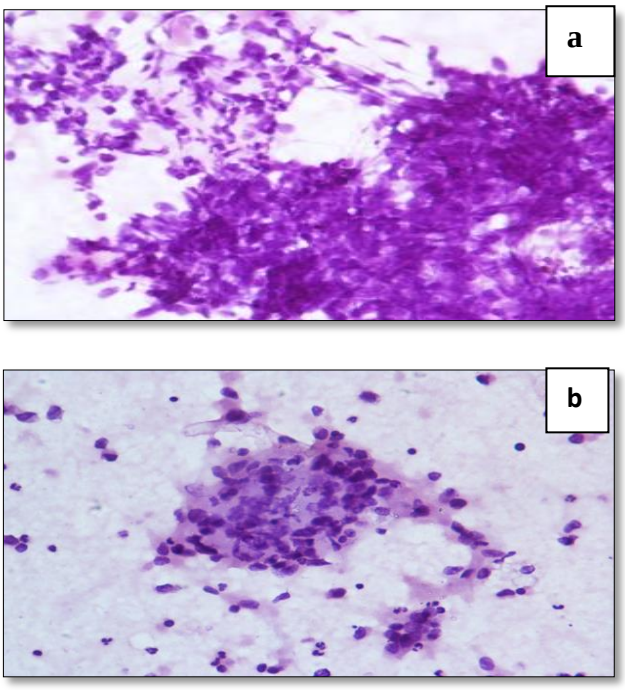

Figure 6a and b: Cells are having scanty cytoplasm and round to oval hyperchromatic nucleus.

Following the diagnosis, surgical excision was done with postoperative radiotherapy.

DISCUSSION

Parapharyngeal space is in the shape of inverted pyramid. The base is formed by greater wing of sphenoid at skull base. The apex is at level of greater cornu of hyoid bone. Medial wall is composed of superior constrictor muscle. Prevertebral fascia forms posterior wall. Lateral wall from antero-posterior direction is formed by medial pterygoid muscle, vertical ramus of mandible, deep lobe of parotid gland and posterior belly of digastric muscle respectively. Parapharyngeal space is divided into prestyloid and poststyloid spaces. Because base, lateral and posterior walls of parapharyngeal space are bony, tumor tends to grow medially. Hence, most common physical sign is medial and inferior displacement of soft palate and tonsillar fossa.

The histopathology of parapharyngeal tumours is diverse. Most common lesions are pleomorphic adenoma.[4] They may arise from deep lobe of parotid. Next most common group of tumors is neurogenous tumors. These include schwannomas, ganglioneuroma, neurofibromas and neuroblastomas. Other tumors are lymphomas, fibrosarcomas, lipoma, hemangiomas, teratomas, undifferentiated carcinomas or metastatic lesions. Undifferentiated carcinomas may arise from deep lobe of parotid. This is a rare salivary gland tumor, comprising 3.2% of primary epithelial tumors of parotid tumors. It is seen predominantly in middle age.[5] There are 2 varieties: small cell type being twice as common as large cell type. This tumor is very aggressive and requires radical surgery followed by postoperative radiotherapy. Five year survival rate is 20-30%.[6]

The standard radiological investigations include computed tomography (CT scan), magnetic resonance imaging (MRI), and angiography and ballon occlusion of internal carotid artery. CT scan and MRI were employed to assess the extent of lesion and plan surgical approach. MRI is considered as superior modality owing to advantages like specific signal intensity characteristics, delineation of fat planes and ability to reveal carotid attenuation by tumours.[7] Surgery is the mainstay of treatment in parapharyngeal tumors. The conventional surgical approaches are trans-oral, trans-parotid, trans-cervical with or without mandibulectomy and midline trans-mandibular -oropharyngeal approaches.[8] Out of them; trans-cervical and trans-parotid are commonly used. The later approach can lead to temporary facial weakness and Frey's syndrome.[9,10]

CONCLUSION

To conclude, parapharyngeal space tumours are rare entity. Undifferentiated carcinomas are even more rarely encountered in parapharyngeal space. Since these tumours are very aggressive in nature with 5 year survival rate of 20-30%, so, radical surgery with postoperative radiotherapy is the main modality of treatment.

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