Unusual Tumors of the Palate

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Abstract

Neoplasms of the palate are rare. These are included in cancers of the oral cavity and pharynx. Two cases of different neoplastic conditions constituting an uncommon group of tumors of the palate, namely, primary Chondrosarcoma and Ganglioneuroblastoma are presented. In the first case, an adult female with a swelling in the hard palate of 18 months duration was diagnosed as chondrosarcoma of the hard palate on light microscopy and was confirmed by immunohistochemistry. In the second case, a female *child with a soft palate mass which was present since birth was diagnosed as ganglioneuroblastoma on light microscopy.*

Key Words: *Palate, tumors, chondrosarcoma, ganglioneuroblastoma*

Introduction

The palate is divided anatomically into the hard palate (part of the oral cavity) and the soft palate (part of the oropharynx)[2]. Neoplasms of the palate, which are included in cancers of the oral cavity and pharynx, have the highest prevalence rate in India[2]. Cancer of the soft palate accounts for approximately 2% of head and neck malignancies[2,3]. Tumors that commonly occur in the region of the palate are squamous cell carcinomas and less commonly the non-squamous malignancies including minor salivary gland tumors, sarcomas and others[2]. Two cases of palatal tumors which are very unusual at this site are presented, namely, primary chondrosarcoma of hard palate and Ganglioneuroblastoma of the soft palate.

Case No. 1

A 30 year old female presented with a painless, firm swelling over the hard palate, measuring 3x2 cm, of 18 months duration, (Fig. 1). Cut surface of the mass was homogenous and cartilage like in appearance. CT scan revealed an extensive heterogenous lesion causing destruction of the maxilla (Fig. 2). Microscopy of H & E stained sections from the mass showed hypercellularity with hyperplastic chondrocytes in diffuse sheets, present in a pale chondroid background (Fig. 3).

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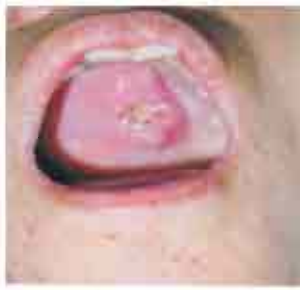

Fig 1: Case 1: Gross, Chondrosarcoma, 3x2 cm firm nontender swelling in the hard palate.

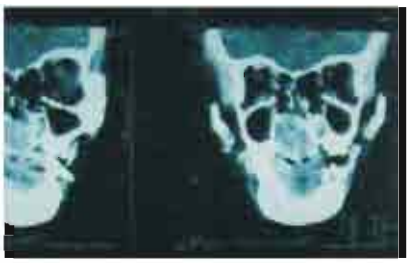

Fig 2: Case 1: CECT: Extensive heterogenous lesion destroying the maxilla

Fig 3: Case 1: Atypical, chondrocytes in lacunae with pale chondroid background (H & E x 400)

Many of them showed atypical nuclei and vacuolated cytoplasm. Few scattered atypical multinucleated giant cells were also seen. Immunohistochemistry revealed S-100 protein positivity, which helped to confirm the diagnosis of chondrosarcoma.

Case No. 2

An 18 month old female presented with a painless, well-defined mass measuring 2x1 cm in the soft palate, present since birth. Cut surface of the mass was gray-white with solid and cystic areas. CT scan revealed a heterogenous soft tissue lesion arising from the palate. The H&E stained sections of the mass on microscopy, showed hypercellular areas of primitive neuronal cells in a schwannian stroma with admixed scattered ganglion cells (Fig. 4,5,7). Focal areas of neuroblastic cells forming rosettes (Fig. 6) and also palisading of nuclei were seen. Hence, the diagnosis of Ganglioneuroblastoma was made.

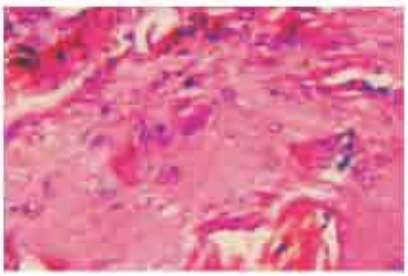

Fig 4 : Case 2 : Highly cellular area of neuronal cells in schwannian stroma (H & E X 400)

Fig 5: Case 2: Highly cellular area showing blastemmal cells with large round vesicular nuclei (H&E X 1000)

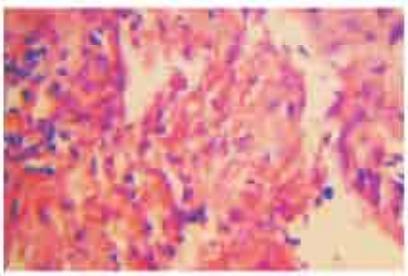

Fig 6 : Case 2 : Neuroblastic cells with indistinct cell borders forming rosettes (H & E x 1000)

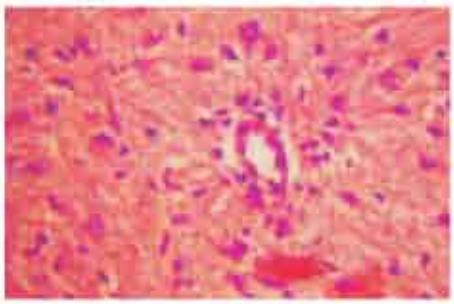

Fig 7 : Ganglion cell with neuroblastic cells in a schwannian stroma (H & E x 400)

Discussion

Case No. 1

Chondrosarcomas are a heterogenous group of malignant tumors arising from cartilage cells that tend to maintain their essentially cartilaginous nature throughout their evolution[3]. Although most chondrosarcomatous tumors arise from cartilaginous or bony structures, they may also develop in soft tissues in which cartilage is normally not found.3 Chondrosarcomas are commonly seen in the pelvis, femur, humerus and other long bones[4]. They account for approximately 11-25 % of all primary sarcomas of the bone. A review of literature reveals that chondrosarcomas of the craniofacial region is extremely rare. Less than 10% of all cases of chondrosarcomas involve the craniofacial region, accounting for less than 2% of all head and neck tumors[3]. In the head and neck region, most chondrosarcomas occur in the maxilla, others being found in descending order of frequency in the body of the mandible, the ramus, the nasal septum and the paranasal sinuses[4].

Chondrosarcomas of the head and neck have been reported in patients ranging in age from 17 months to 75 years with peak age of incidence being the third to the sixth decade[4]. Craniofacial chondrosarcomas most commonly present as a painless mass/swelling,[3] as was in the case being discussed. Diagnosis can only be established by histopathological examination.[5] Recent studies suggest that the pathogenesis of chondrosarcomas may involve mutational inactivation of the p16, Rb and p53 tumor suppressor genes[3,6]. Although local spread may occur commonly, distant metastasis is rare, and if present, most commonly involves the lungs[4].

Surgery is the main stay of treatment for patients with chondrosarcomas of the head and neck[3].

Chondrosarcomas are radioresistant lesions.[5] Grading of this tumor is based on cellularity and nuclear enlargement and irregularity, with scores of [1,2] and [3] (low, intermediate and high grade respectively)[3]. The prognosis is generally good for low and intermediate grade chondrosarcomas. Prognosis is also related to the location of the tumor, and the adequacy of primary surgical resection[3]. The overall 5 year disease free survival for low grade chondrosarcomas after complete resection ranges between 54 to 77%[3]. However, when localized to the facial bones, as in this case, they follow a more aggressive pattern as opposed to when localized in long bones, as the former has a greater rate of growth, recurrence and metastasis[7]. Differential diagnosis of chondrosarcomas of the palate mainly include chondroma, chondroblastic osteosarcoma, chondromyxoid fibroma, chordoma and odontogenic tumors[8].

Case No. 2

Neuroblastoma, ganglioneuroma and ganglioneuroblastoma (GNB) are collectively known as neuroblastic tumors. Neuroblastic tumors are tumors of the sympathetic nervous system. They originate from neural crest sympathogonia, which are completely undifferentiated cells of the sympathetic nervous system[10].

Neuroblastoma, ganglioneuroblastoma and ganglioneuroma can be conceptualized as three maturational manifestations of a common neoplasm[4]. Neuroblastoma (NB), being the least differentiated, resembles the fetal adrenal medulla and is made up of primitive neuroblasts. Ganglioneuroblastoma which is considered to be a differentiating neuroblastoma, has primitive neuroblasts along with maturing ganglion cells. Ganglioneuroma, a fully differentiated tumor, is characterized by a mixture of mature Schwann cells and ganglion cells. Neuroblastoma and ganglioneuroblastoma, are both considered malignant and hence are discussed together[4].

Neuroblastoma is the most common malignancy in infants under the age of 1 year. It occurs at a rate of about 1/ 10000 live births[4]. It accounts for 10-12 % of all malignant tumors, preceded in frequency only by leukaemias and brain tumors[4]. About one-fourth of all neuroblastomas are congenital.

The distribution of this tumor generally follows the distribution of the sympathetic ganglia, hence they are found in the paramidline position at any point between the base of the skull and the pelvis, in addition to the adrenal medulla and organ of Zuckerkandl[4]. Locations such as urinary bladder, bowel wall, abdominal wall and gall bladder are considered unusual[1]. Soft palate is an extremely rare site for this tumor. An extensive search of literature did not yield references regarding occurrence of this tumor in the soft palate, though references regarding its occurrence in the hard palate are present. Presenting signs and symptoms vary depending on age of the patient, location of the mass and presence or absence of associated clinical syndromes. In half the cases, a nodular fixed mass extending across the midline can be palpated[4]. About one-third of neonates with neuroblastoma present with blue-red cutaneous metastases which have been likened to blueberries (blueberry muffin baby). About one-fifth of the patients have hypertension which remits with tumor removal. A relatively rare presentation, usually associated with good prognosis, is the myoclonus-opsoclonus syndrome[4].

About 80-90 % of patients with neuroblastoma have elevated levels of catecholamines and their metabolites (VMA, HVA) in their urine[4]. HVA tends to be secreted by more mature and differentiated tumors and VMA is usually a product of less differentiated tumors[10]. Catecholamine toxicity however rarely results[10]. Ferritin can be detected in the serum of patients with active disease and is also used as a prognostic indicator[4]. These tumors are staged according to the international neuroblastoma staging system (INSS) developed in the mid 1980s.11 Patients with either a low or intermediate grade of tumor have relatively good prognosis.

Non specific enolase (NSE) is the most sensitive but also the least specific immunohistochemical marker for neuroblastoma and is identified with increasing intensity in differentiating tumors, such as ganglioneuroblastomas and ganglioneuromas. However, as it is not very specific, NSE cannot be used alone for the differential diagnosis[4].

Ganglioneuroblastoma of the palate needs to be differentiated from neuroblastomas, melanotic neuroectodermal tumors, neurofibromas, schwannomas and ameloblastic tumors[10].

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