Original Article

A Study of Rare Cases of Head and Neck Region in a Tertiary Care Centre in Last Three Years

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Introduction

Head and neck cancers include a wide range of uncommon tumours that occasionally develop aggressive biological behavior. Within the head and neck region, there are a wide number of sites and subsites where such primary tumours can develop, that differ accordingly in epidemiology, presentation and management modalities.

This article studied five such cases of uncommon tumours in the head and neck region presenting over a period of three years in the hospital. It included their clinical, radiological and histopathological features that created a diagnostic dilemma but were resolved by correlations between the clinicians and pathologists. The five unusual cases included:

A) A case of a 22 year old male patient presented in June 2015 with left sided nasal obstruction for 1 year, diagnosed as Maxillary Osteosarcoma.

Osteosarcoma is a highly aggressive malignant bone tumor almost exclusively involving the metaphysis of long bones of the appendicular skeleton, characterized by neoplastic osteoblasts producing osteoid material.¹ The involvement of nonlong bones such as the skull, jaw, and spine are extremely rare. Only 6% to 10% cases occur in the craniofacial region.²

It is primarily a tumour of adolescent and young age, as 60% of patients are younger than 25 years of age.³ Patients with osteosarcoma commonly present with bony mass, pain, and nerve compression symptoms. Nasal obstruction and discharge mimicking sinusitis is extremely rare as a presenting feature.⁴

B) A case of a 23 year old male patient presented in June 2015 with swelling in the right lateral border of tongue and was diagnosed as Lingual Schwannoma.

Schwannoma (neurilemmoma) is a solitary, benign, encapsulated, slow growing tumour originating from Schwann cells of peripheral sensory or motor nerve.⁵ Though the overall incidence of Schwannoma is 25-45% in head
& neck region, only 1% is seen in oral cavity, among which tongue is the most common site. Because of its low incidence and absence of definite signs and symptoms, it is often not included in differential diagnosis of tongue swelling. Histopathological findings are sufficient for diagnosis.6

C) A case of a 24 year old male patient presented in June 2016 with throat discomfort, had a polypoidal growth on left tonsil and was diagnosed as Tonsillar Lipoma. Lipomas are common benign mesenchymal tumors that can arise wherever fat can normally be present. They tend to be slow growing with a peak incidence between 5th to 6th decade of life7,9. Over 10% lipomas arise in head and neck region, usually in the immediate subcutaneous tissue, among which only 1-4% are noted within oral cavity.7,8 In oral cavity; buccal mucosa, mouth and tongue are the frequent sites.7 Lipomas originating from the tonsil are unusual. Since tonsillar lipomas do not cause severe symptoms; they are diagnosed incidentally and treated surgically only for cosmetic reasons 7–9.

D) A case of a 9 years old male presented in March 2018 with nasal discharge and was diagnosed as Nasal Glioma
Most cases of Nasal gliomas present in newborns, with more than 90% of the cases diagnosed by age of 2 years. The lesion is situated externally on or near the bridge of the nose in 60% of the cases, within the nasal cavity in 30% of the cases or both in the rest 10% cases, where a defect in nasal bones allow for the communication.10

E) A case of a 52 year male presented in May 2018 with a mass in the medial aspect of left orbit for three months and was diagnosed as Epithelioid Haemangioma with eosinophilia.
Epithelioid hemangioma, also known as angiolymphoid hyperplasia with eosinophilia, has been a controversial lesion regarding it being a reactive proliferation or a neoplastic process involving the mesenchymal element, mostly around the ear and temporal region. It most frequently occurs in the third to fifth decade of life without any gender predilection.11

Material and Methods
This study was conducted in the Department of Pathology in collaboration with the Department of Surgery. Time period of this study was from June 2015 to May 2018. The study was a cross sectional observational study.
The study population comprised patients of all age groups who presented with symptoms representing mass lesions in the head and neck region and were examined clinically in the tertiary care hospital. Ethical clearance and consent was taken for the cases. After prior investigations like radiography, operative procedures were done in each case and the specimen received was studied for histopathology. After correlating the histomorphological features in each case with the clinical findings and incidence of such tumours in unusual locations, the histopathological diagnoses of such cases in these locations of head and neck were found to be quite rare.
The case history for each case was collected and noted as follows:

Maxillary Osteosarcoma
A 22-year-old male patient presented in the ENT OPD with the complaints of left-sided nasal obstruction for 1 year. He was provisionally diagnosed with chronic sinusitis, but his symptom was not relieved by medications. Over time, a slowly growing painless swelling appeared over his left cheek. On physical examination, a 5 ×3 cm ill-defined, hard, nontender swelling was palpable over the left maxillary region. No cervical lymphadenopathy was detected. Examination of other systems was unremarkable. Complete blood count, chest X-ray, and abdominal ultrasonography were found to be within normal limits.
Towne’s (OM) view plain X-ray of paranasal sinuses revealed a densely calcified and ossified mass in left maxillary antrum. Plain and contrast-enhanced computerized tomography (CT) scan of paranasal sinuses was done, with 5 mm sections both in the axial and coronal planes. Left maxillary antrum showed expansile, ill-defined, mixed radiopaque-radiolucent lesion, invading the cortical bone and adjacent soft tissues resulting in a “sunburst appearance” of periosteum. The lesion extended into frontosuperior margin of left orbit and ethmoid air spaces occluding the left nasal cavity. The mass presented dense amorphous ossifications and showed the heterogeneous uptake of contrast material [Figures 1 and 2].

**Lingual Schwannoma**
A 23 year old male presented with a swelling at right lateral border of tongue for three months without any complain of pain, difficulty in deglutition, chewing and phonation. On local examination, a small, firm, nontender, submucosal swelling was noted. Examination of oral cavity was unremarkable. Due to frequent history of tongue bite, lesion was provisionally diagnosed as traumatic fibroma.

The patient underwent transoral resection. Surgery was uneventful and the specimen was sent for histopathological examination.

**Tonsillar Lipoma**
A 24 year male, attended the otolaryngology clinic with complain of throat discomfort which he described as a feeling of something stuck in his throat. There was no history of pain, fever and change of voice quality. The patient had no contributory past medical history.

On clinical examination, a well-defined, small, mobile, non-tender, intraglandular polyloid growth was noted, arising from the upper pole of left sided tonsil, measuring 1x1 centimeters. Elective excision of the polyp was performed and the excised tissue was sent for pathological examination.

**Nasal Glioma**
A 9 year male presented to the ENT OPD with a firm subcutaneous, non compressible nodule at the bridge of the nose along with nasal discharge for one month. The nasal discharge was detected as cerebrospinal fluid leak.

On radiological finding, an expansile mass was noted at the same site. After excision, the mass was sent for histopathological examination.

**Epithelioid Hemangioma with Eosinophilia**
A 52 year male presented to the ophthalmology clinic with a mass in the medial aspect of left orbit. Vision was normal in the left eye whereas the perception of light was negative in the right eye due to prior trauma. The C.T. scan of orbit revealed a space occupying lesion in the upper medial aspect of left orbit. The patient underwent an excision of the mass and the specimen was sent to the Department of Pathology.

The specimen of all the above mentioned cases were received and grossed as per standard protocol. After processing the tissue sections, H&E staining was done.

**Results**
The gross pathological features as well as the histopathological features of each case was noted as follows:

**Maxillary Osteosarcoma**
Biopsy of the growth revealed histopathological features compatible with osteosarcoma. Left hemimaxillectomy was performed including a wide excision of the tumour, and the specimen was sent for histopathological examination. On gross examination, a hard bony growth was found in the maxilla, measuring 5 × 5 × 3 cm. The cross section showed variegated appearance [Figure 3]. Microscopic examinations showed abundant eosinophilic lace-like osteoid materials lined by highly pleomorphic malignant osteoblasts, confirming the diagnosis of osteosarcoma [Figure 4]. Sections from orbital plate, skin over maxilla, and posterior and lateral margins also revealed the presence of tumor. The patient was referred to the
Department of Radiotherapy and was on close follow-up then.

**Lingual Schwannoma**
Grossly the resected specimen was a single, greyish-white tissue, measuring 1.2 x 0.8 x 0.5 cm. Cut section was solid, white and homogenous (Figure -5).

Microscopy revealed a well encapsulated mass (Figure-6) having cellular area comprising of bland spindle cells (Antoni A) and loose myxoid area (Antoni B) (Figure -7). Verocaybody and thick hyalinised blood vessels are also seen. Histopathological diagnosis of Schwannoma was made and confirmed by immunohistochemistry (S100).

**Tonsillar Lipoma**
Grossly, the tissue was described as thinly encapsulated, polypoid mass, measuring 1.2 cm x1cm x 0.5 cm with a, soft, yellowish, homogenous cut surface.

Microscopically, histological sections of the tissue revealed a well circumscribed, encapsulated tumor composed of mature adipocytes with no evidence of atypia (Figure 8).The cells were arranged in lobules. A rim of lymphoid tissue remained at the periphery of the lesion. The diagnosis of lipoma was made based on the histopathological features (Figure 9). Routine follow up revealed no residual abnormality.

**Nasal Glioma**
A partially skin covered polypoidal mass was noted in macroscopy, measuring 2.5 cm in maximum dimension and having a soft. grey tan cut surface.

On microscopic examination, nests and islands of glial tissue was noted, separated by bands of fibrous connective tissue. Mitoses was absent in the sections examined.

**Epithelioid Hemangioendothelioma with Eosinophilia**
On macroscopy, a nodular mass was noted, measuring 1cm in maximum dimension. On microscopy, proliferation of vascular channels was noted, along with dense inflammatory cell infiltration, comprising lymphoid cells and rich collections of eosinophils. The vascular spaces were lined by plump-appearing (epithelioid) endothelial cells with mild pleomorphism, hyperchromatic nuclei, copious eosinophilic cytoplasm, and inconspicuous nucleoli.
Figure 3: A variegated bony growth found in the maxilla on gross examination (inset, cross section).

Figure 4: Photomicrograph showing abundant eosinophilic, lace-like, osteoid material lined by highly pleomorphic malignant osteoblasts confirming the diagnosis of osteosarcoma (H and E, 400x).

Figure 5: gross photograph of cut section of tissue showing whitish, solid tumor

Figure 6: low power view showing tumor mass composed of hypocellular and hypercellular areas (H&E, 40X)

Figure 7: high power view showing Antoni A areas having bland spindle cells forming verocay body (H&E, 100X)

Figure 8: Photomicrograph of polypoid tonsillar mass showing lobules of mature adipocyte beneath the lymphoid follicles. (H&E, x100)
Discussion

The individual cases were discussed as follows:

**Maxillary Osteosarcoma**

Osteosarcoma is the most common primary bone tumor, but only 6% to 10% cases occur in the craniofacial region. Among the cases of craniofacial region, mandible is a comparatively common site, followed by maxilla and skull. When compared with other regions, the craniofacial osteosarcomas are less aggressive and supposed to have local invasion rather than distal metastasis.12

Patients with craniofacial osteosarcoma commonly present with bony mass, pain, and nerve compression symptoms. Sinusitis-like features as present in the index case are extremely rare in clinical literatures. The risk factors for this tumor are preexisting bone diseases or the genetic defects, which are similar to the risk factors for osteosarcoma in other regions. The common preexisting bone diseases that can lead to osteosarcoma are Paget’s disease, fibrous dysplasia, chronic osteomyelitis, giant cell tumor.13 The patient in this study did not reveal any of these conditions.

Radiological findings of the craniofacial lesions are nonspecific. They can present as an osteosclerotic/mixed/osteolytic lesion. CT scan plays an important role, as it can identify the exact extent of the mass, bony erosion, soft-tissue infiltration, and degree of ossification.14 The CT findings in this case showed a mixed density lesion, which eroded bone and adjacent soft tissue and invaded the orbit. Reported craniofacial cases show an equal incidence of osteoblastic, chondroblastic, and fibroblastic types.15 In this case, the microscopic features were of osteoblastic osteosarcoma.

The ideal treatment for this tumor is complete resection. Total maxillectomy is the recommended surgical procedure. Excision with clear surgical margin is prognostically better and will result in a longer survival.16 Adjuvant treatments must be considered for total complete eradication of the tumor and to prevent recurrence.17 In this case, the
skin over maxilla and the posterior and lateral margins were involved by the tumor.

**Lingual Schwannoma**

Only 1% of head and neck schwannomas occur in oral cavity, of which the most common site is lateral border of tongue (36.3%), followed by base (24.2%), tip (21.2%) and ventral surface (15.1%) 18. It is usually a solitary tumor occurring in 2nd to 3rd decade of life without any gender predilection 19. The risk of malignant transformation is very low (8-10%) 20. Lingual schwannomas are usually asymptomatic when the average size is less than 18.2mm and symptomatic when more than 33mm 21.

Clinically, it can be confused with neurofibroma, traumatic fibroma, lipoma, leiomyoma 22. MRI can be used to locate the extension of the tumor which helps in surgical planning. Complete excision is the treatment of choice. Histopathology is almost always confirmatory which can be further corroborated by immunoreactivity for S100. Recurrence after complete excision is extremely rare 24.

**Tonsillar Lipoma**

Tonsillar tissue histology consists of a combination of centrally-localized lymphoid tissue and epithelial tissue composed of squamous cells covering crypts and all surfaces 9. There is no adipose tissue in this site, therefore lipomatous tumor incidence is quite low in the tonsil 25-27. Lipomas are composed of mature adipocytes and are frequently encountered benign mesenchymal tumors, because they can originate anywhere within the body where adipose tissue is located 28, 29. Generally, their prevalence does not differ with gender, although a male predilection has been recorded 30. These tumours can be seen in the buccal sulcus, tongue, floor of mouth and lips more, than the lower pole of the tonsil and hypopharyngeal walls 9. In addition to squamous papilloma, the other benign tumors such as adenoma, lipoma, chondroma, hamartoma and teratoma should be considered as the differential diagnosis of benign tonsillar tumors 27-30.

The etiology is unknown. However, it is thought that trauma may trigger proliferation of fatty tissue and cause lipomas. Clinically, tonsillar lipomas are slow growing painless masses and patients commonly present with a well-circumscribed mass that have been developing for several years, but they may manifest symptoms such as voice change, dysphagia, soreness, excessive salivation or foreign body sensation as in our reported case. Furthermore, when they reach an important size, these tumors may cause respiratory obstruction. Dereköy et al 26 defined a case of tonsil lipoma that was 3.6 centimeters in length and caused dyspnea, hypoxemia and respiratory acidosis.

Although MRI and CT scans are very useful in the clinical diagnosis, the histopathology remains the gold standard in the diagnosis of lipomas. On histological analysis, lipomas can be classified as simple/classic lipoma, fibrolipoma or angiofibrolipoma based on the varying amount of fibrous tissue, capillaries and/or lymphatics. Differential diagnosis includes other benign tumors of the tonsils such as papilloma, which represent the most common neoplasm of the tonsils 30. Although extremely rare, malignant change in lipoma was described. Saddik et al reported one case of an under-diagnosed liposarcoma of the tonsillar fossa. 26 Surgical excision is the usual mode of treatment in symptomatic cases 25.

**Nasal Glioma**

Nasal gliomas include both nasal encephaloceles and nasal glial heterotopias. Both are generally considered to be heterotopic tissue of central nervous system that has been displaced anteriorly. Characteristically, such heterotopic tissues are located in the nasal region, but similar tissues can also be noted in the scalp and other skull lesions. Nasal encephaloceles represent herniation of brain tissue and leptomeninges through a bony defect in the skull. It is thus typically identified
with association of a discernible cranial bone defect and is often secondary to infection, trauma or surgery. Nasal glial heterotopias on the other hand, are congenital malformations of displaced normal, mature glial tissue and may not have a cranial bony defect. Based on location, they can be either extra nasal (60%), presenting as a subcutaneous lesion on the bridge of the nose, or intranasal (30%), presenting interiorly in the superior nasal cavity, or mixed (10%).

On microscopy, the glial tissue appears similar to gliosis. The fibrous connective tissue is blended with the glial tissue. Fibrosis frequently obliterates the glial tissue. Within the glial tissue, gemistocytes may be noted but neurons are uncommon. encephalocele shows glial degeneration. 31

Ancillary investigations include histochemistry stains like trichrome stain, staining glial tissue bright red and fibrosis blue. Immunohistochemistry for glial tissue is positive with S100 and GFAP.

Epithelioid Haemangioma (EH) with Eosinophilia

EH often presents as a subcutaneous proliferation with a predilection for the external ear as well as other head and neck sites, including the scalp and forehead. 32,33,34 EH most frequently occurs in the third to fifth decade of life; no gender predilection is evident. The symptoms include pruritus and bleeding following scratching. Regional lymphadenopathy and peripheral eosinophilia are uncommon. Human herpesvirus-8 (HHV8) has not been identified in association with EH. 36, 37, 40

Local surgical excision or desiccation is the treatment of choice, and these are curative. Recurrences are occasionally seen. Medical regimens, including intralesional or systemic steroids, have been used with some success in treating the symptoms, but they have not been curative.

EH is characterized by single or multiple, pink to red-brown indurated cutaneous papules or subcutaneous nodules. These lesions measure from a few millimeters to 1 cm in diameter. Clusters of papules may coalesce to form large, plaque-like lesions. 32,33 Histologically, EH is characterized by a nodular vascular proliferation that is accompanied by a variably dense lymphoid infiltrate that is rich in eosinophils. The process is circumscribed but not encapsulated, and it may involve the subcutis, dermis, or both. The vascular component varies in size from capillary to medium-sized arteries and veins, and the vascular spaces are lined by plump-appearing (epithelioid) endothelial cells with mild pleomorphism, hyperchromatic nuclei, copious eosinophilic cytoplasm, and inconspicuous nucleoli. 32,35,39 Frequently, the endothelial cells protrude into the vessel lumen in a “hobnail” fashion, creating a cobblestone-like appearance. 38

The vessels vary from irregular, poorly canalized, thin-walled spaces to rounded, well-formed vessels with thickened walls. Origin from a small artery or vein is common but may be dependent on adequate sampling. It is common for the entire lesion to be intravascular. 36, 37 Surrounding the vascular proliferation, a prominent inflammatory infiltrate is commonly seen including mature lymphocytes, histiocytes, and eosinophils. The endothelial cells in EH are immunoreactive for CD31 and to a lesser extent CD34. Staining for glucose transporter protein 1 (GLUT-1) is typically absent. Rarely, cytokeratin reactivity has been reported. 41, 42

Conclusion

In conclusion, it can be said that all the cases discussed in this article is notable due to its rarity regarding the location and presentation. Osteosarcoma of craniofacial region generally presents in the advanced stage in the tertiary-care hospital, mainly, because of rarity of occurrence and lack of awareness among the treating physicians. Our case is unique on account of its rare presenting features and unusual site of occurrence.

Schwannoma is very rare in oral cavity especially in tongue. It should be considered as a differential
diagnosis while dealing with tongue mass. Our case is unique due to rarity of its location. The case of tonsillar lipoma was reported to raise the level of awareness of this benign lesion in the tonsil and to emphasize the importance of appropriate histopathological evaluation. Nasal glioma is a controversial lesion. It has negligible malignant potential, thus the term “glioma” in the clinical, surgical or radiological literature is misleading. Epithelioid hemangioma (EH), also known as angiolymphoid hyperplasia with eosinophilia (ALHE), has been a controversial lesion with regard to its classification (reactive proliferation or neoplastic process) and its relationship to Kimura disease. Although a reactive etiology especially secondary to trauma has been proposed, a neoplastic origin is favored. EH is considered to represent the benign end of the spectrum of vascular tumors characterized by epithelioid endothelial cells, many of which are rich in lymphocytes and eosinophils. The malignant end of the spectrum includes epithelioid haemangioendothelioma and epithelioid angiosarcoma.

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