Case Report

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Dentistry Section

Primary Malignant Mixed Germ Cell Tumour with Squamous Cell Carcinoma of the Mandible; A Rare Entity

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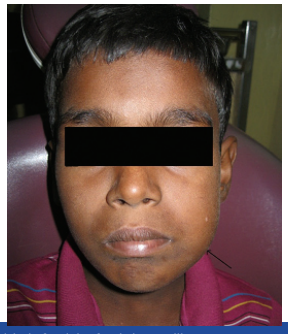
ABSTRACT

Germ cell Tumours (GCT) are neoplasm derived from germ cells. GCT usually occurs inside the gonads. Extragonadal GCT's are rare. Most common GCT associated with head and neck region are the teratomas. Of the few teratomas found in the head and neck, malignant transformation of a teratomatous element is very uncommon, and primary bone involvement within the head and neck is even rare. We present a case of primary malignant mixed germ cell Tumour involving the mandible, the present case presented malignant transformation of the epithelial component showing foci of squamous cell carcinoma within the GCT.

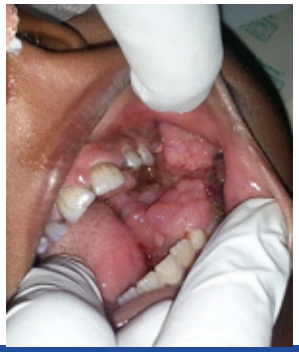
Keywords: Mandible tumour, Extragonadal tumour, Paediatric malignancy, Teratoma

CASE REPORT

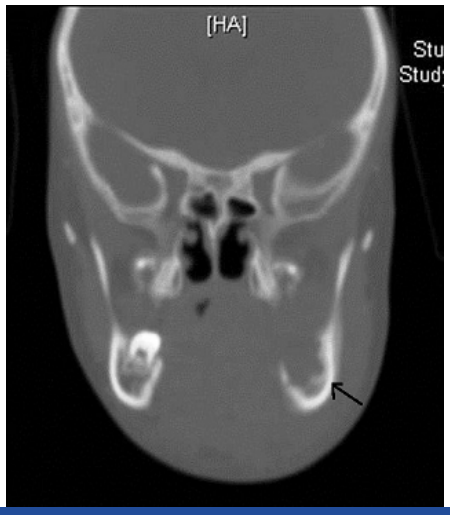
A 10-year-old male presented as an outpatient in the Department of Dental and Oral Surgery, Christian Medical College and Hospital, Vellore; with a pale pink, raised, firm, painless gingivo-buccal growth (approximately 4cm x 4cm) on the left posterior alveolus along with mild left sided facial swelling [Table/Fig-1,2]. The patient's parents first noticed facial swelling approximately two months prior to presentation. The patient has nil significant family history. Patient had normal mouth opening. The growth was sessile and was diffusely adhering to the left posterior alveolus extending to the left gingiva-buccal sulcus. The tongue was freely mobile and floor of the mouth was intact and free of the tumour. There was no significant induration around the lesion. The left submandibular lymph node was palpable but clinically non-significant. Coronal CT and radiographs showed significant bony erosion in the lateral portion of the left alveolus and mandibular body [Table/Fig-3,4].



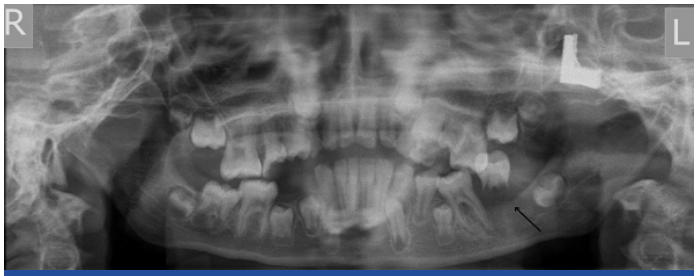
[Table/Fig-1]: Patient with left side facial swelling



[Table/Fig-2]: Intraoral proliferative growth over the left side lower posterior alveolus

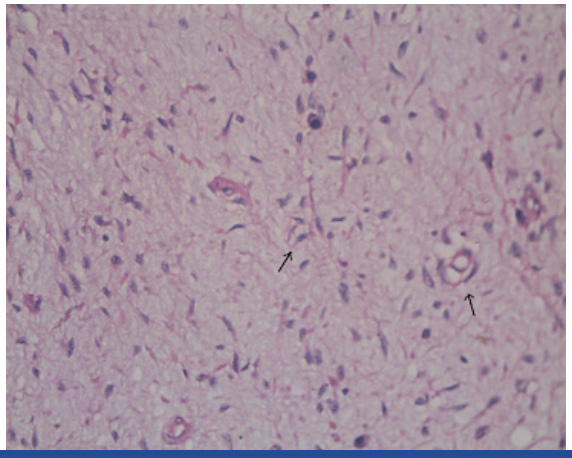


[Table/Fig-3]: Coronal CT showing bony erosion of the left side mandibular alveolous

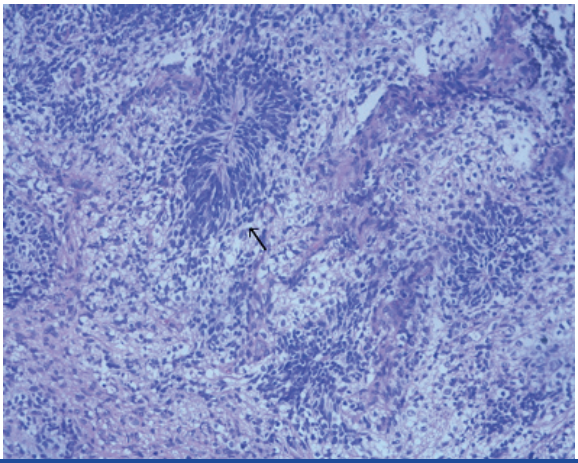


[Table/Fig-4]: Orthopantomogram showing involvement of the left side body of mandible

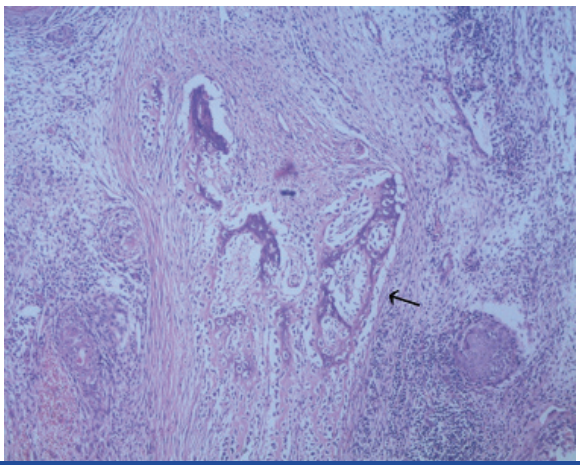
Based on clinical assessment and aggressiveness of the tumour differential diagnosis were varied and primarily included germ cell Tumour/teratoma, sarcomas inclusive of Rhabdomyosarcoma, Ewings/Neuroectodermal Tumour, Histiocytosis. Also, possibility of Non- Hodgkin's lymphoma was considered. Incisional biopsy of the lesion was performed. Microscopically, on histopathologic staining the tumour was composed of heterogenous components including immature and mature neuroepithelium [Table/Fig-5,6], primitive mesenchyme with bone [Table/Fig-7], nodules of cartilage [Table/Fig-8], squamous morules [Table/Fig-9], foci of glandular differentiation [Table/Fig-10] lined by polygonal cells with mitotically active, hyperchromatic nuclei and variable amounts of cytoplasm



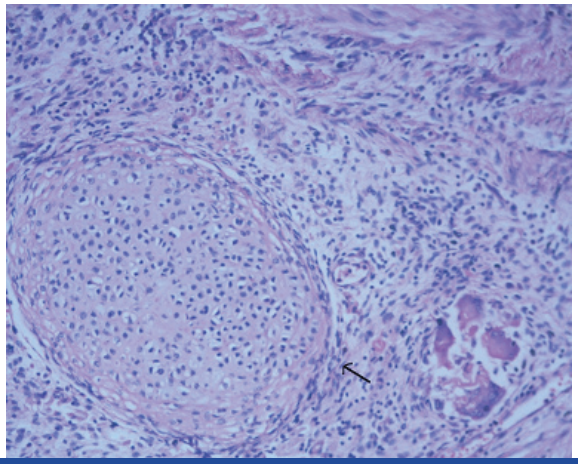
[Table/Fig-5]: H&E stain (20X) Images show parts of a tumour composed of mature neural tissue



[Table/Fig-6]: H&E stain (20X) Image shows part of a tumour composed immature neural component

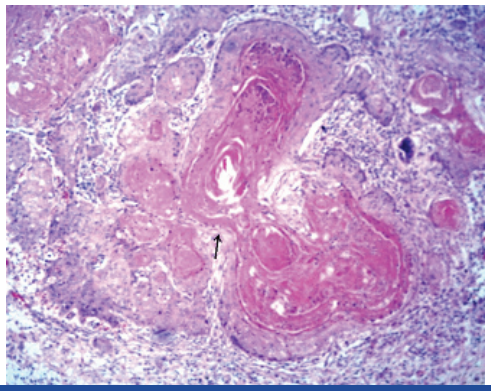


[Table/Fig-7]: H&E stain (20X) Image shows primitive mesenchyme with bone

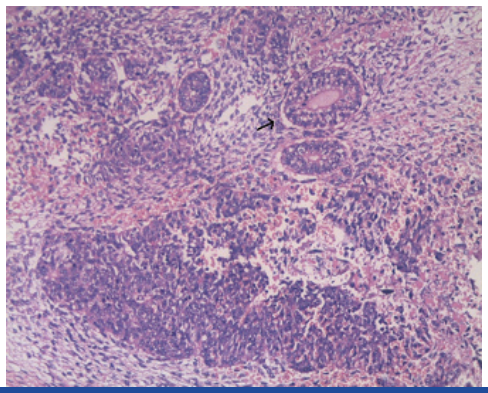


[Table/Fig-8]: H&E stain (40x) Image shows nodules of cartilage

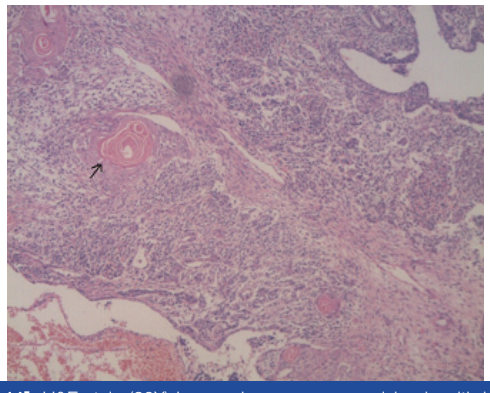
set in a cellular fibroblastic stroma in areas. There were squamous islands with keratin pearl formation, suggesting foci of squamous cell carcinoma [Table/Fig-11] henceforth a diagnosis of Primary malignant mixed germ cell Tumour with squamous cell carcinoma of the mandible was rendered. Serum AFP was 1.30IU/ml (normal <5.5IU/ml), beta-HCG was 0.675mIU/ml (normal <5mIU/ml). Following two cycles of cisplatinum chemotherapy the residual tumour was excised with prophylactic selective neck dissection. The surgical pathology was in correlation with malignant GCT. The neck nodes were free of malignancy. Patient had an uneventful recovery, and has been kept under follow-up. At last visit patient has two years disease free follow-up [Table/Fig-12,13].



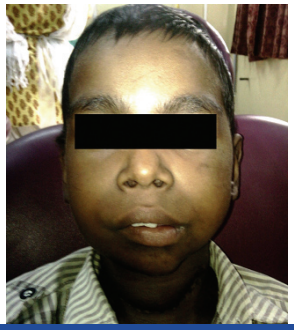
[Table/Fig-9]: H&E stain (40X) Image shows squamous morules



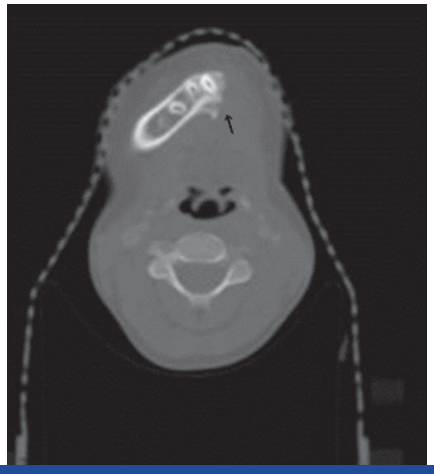
[Table/Fig-10]: H&E stain (40X) Image shows foci of glandular differentiation



[Table/Fig-11]: H&E stain (20X) Image shows squamous islands with keratin pearl formation



[Table/Fig-12]: Two year Postoperative follow up profile picture



[Table/Fig-13]: Two Year follow–up axial CT showing normal resected area

DISCUSSION

Malignant germ cell tumours (GCT) represents 3% of neoplasms in the paediatric population. In young children, the majority of these tumours affect extragonadal sites, principally the sacrococcygeal region. Adolescents and adults usually present with a gonadal primary [1]. GCT occurring outside the testes or ovaries are termed extragonadal. In children, two thirds of germ cell neoplasms arise in extragonadal sites, and 40% of these are in the sacrococcygeal region.[2] The head and neck is rarely represented, accounting for only 6% of GCT cases [3]. Reported sites are usually of soft tissue and include the oral cavity, pharynx, orbital region, temporal bone, and neck. The tumours are typically midline in location, only rarely occurring laterally. The cervicothyroidal area is the most common region of presentation in the head and neck [4,5]. Two principal theories are posited to explain the aetiogenesis of GCT. The "germ cell theory" denotes malignant transformation occurring within primordial germ cells during the course of their migration along the urogenital ridge. This helps explain the midline propensity, but does not easily account for a laterally placed cephalad tumour. The "embryonic cell theory" suggests that totipotent embryonic (somatic) cells undergo malignant degeneration into yolk sac Tumour [6].

The indexed case presented as primary malignant germ cell Tumour of the mandible. The histology was typical for this Tumour. In literature primary malignant GCT arising from the oral cavity is rarely reported. Sporadic cases reported includes Hudson et al., [7], who described adenocarcinoma arising in a teratoma of the posterior mandible in a 14-year-old girl, Hudson et al., identified adenocarcinoma of the gastrointestinal-type mucosa as the malignant element, in contrast to the indexed case, where the malignancy was squamous cell carcinoma in nature. Further review of the literature showed two similar cases. Kuhn et al., [8] described squamous cell carcinoma arising in a teratoma of the maxilla in a 4-month-old male infant. Cabay et al., identified squamous cell carcinoma arising in a mature cystic teratoma in the posterior mandible in a 2-year-old boy [9]. These reports all document the involvement of the local hard and soft tissues. Magliocca et al., has reported first case of an epidermoid malignancy developing in a teratoid/teratomatous lesion occurring exclusively within the osseous tissue of the mandible of a 12-year-old female [10], in contrast the present case presented in 8-year-old male child with squamous cell carcinoma arising from immature teratomatous lesion of the mandible. Patients with gonadal GCTs can achieve long-term disease-free survival when chemotherapy is combined with expert and judicious resection of residual disease [11]. The overall prognosis for teratomas of the head and neck region (exclusive of brain and spinal cord) is excellent, despite the presence of immature elements; however, tumour-related deaths can result from large unresectable primary Tumours [4]. However, for extragonadal GCT surgery should always be considered for resectable masses following salvage therapies or in chemoresistant disease to maximize chance of cure [12].

CONCLUSION

Malignant primary GCT of mandible is one of the rarest entity and requires prompt multidisciplinary care for therapeutic outcome. Thorough understanding of pathology as well as early diagnosis based on imaging and histopathologic evaluation is important for favourable outcome. However, the prognosis for children with malignant germ cell Tumour still remains unclear and successful management of these patients depend largely on proper diagnosis with early rigorous adjuvant therapy.

REFERENCES

[1] Steinbacher DM, Upton J, Rahbar R, Ferraro NF. Yolk Sac Tumour of the Mandible. *J oral maxillofac surg*. 2008;66(1):151-53.
[2] Kebudi R, Ayan I, Darendeliler E, Agaoglu L, Kinay M, Olgaç V, et al. Non-midline endodermal sinus Tumour in the head and neck region: A case report. *Med Paediatr Oncol*. 1993;21(9):685-89.
[3] Dehner LP, Mills A, Talerman A, Billman GF, Krous HF, Platz CE. Germ cell neoplasms of head and neck soft tissues: a pathologic spectrum of teratomatous and endodermal sinus Tumours. *Hum Pathol*. 1990;21(3):309-18.
[4] Lack EE. Extragonadal germ cell Tumours of the head and neck region: Review of 16 cases. *Hum Pathol*. 1985;16(1):56-54.
[5] Margo CE, Folberg R, Zimmerman LE, Sesterhenn IA, et al. Endodermal sinus Tumour (yolk sac Tumour) of the orbit. *Ophthalmology*. 1983;90(12):1426-32.
[6] Schneider DT, Schuster AE, Fritsch MK, Hu J, Olson T, Lauer S, et al. Multipoint imprinting analysis indicates a common precursor cell for gonadal and nongonadal paediatric germ cell Tumours. *Cancer Research*. 2001;61:7268–76.
[7] Hudson JW, Jaffrey B, Chase DC, Gray J. Malignant teratoma of the mandible. *J Oral Maxillofac Surg*. 1983;41(8):540-43.
[8] Kuhn JJ, Schoem SR, Warnock GR. Squamous cell carcinoma arising in a benign teratoma of the maxilla. *Otolaryngol Head Neck Surg*. 1996;114(3):447-52.
[9] Cabay RJ, Setty S, Schwartz JL, Yao M, Schmidt ML, Gordon SC. Paediatric squamous cell carcinoma arising in an alpha-fetoprotein-producing mature cystic teratoma of the mandible. *Paediatr Blood Cancer*. 2009;52(1):130-32.
[10] Magliocca KR, Stone IE, Bhattacharyya I. Malignant Cystic Teratoid Lesion of the Mandible. *J Oral Maxillofac Surg*. 2012;70(3):636-32.
[11] Shin YS, Kim HJ. Current Management of Testicular Cancer. *Korean J Urol*. 2013;54(1):2–10.
[12] Daneshmand S, Albers P, Fossa SD, Heidenreich A, Kollmannsberger C, Krege S, et al. Contemporary management of postchemotherapy testis cancer. *Eur Urol*. 2012;62(5):867-76.

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