Teratocarcinoma of Maxillary Antrum: A Rare Presentation of Extragonadal Germ Cell Tumor

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Abstract

Germ cell tumour (GCT) is a neoplasm derived from germ cells. Extra gonadal GCTs (EGGCT) are that which origin outside the gonads as a result of errors during development of embryo. The Mediastinum is the most common anatomic site for EGGCTs in adults. Maxillary GCTs are very rare and commonly occurs within the first three decades of life. Seminomatous EGGCTs variety have better prognosis than non seminomatous variety when treated with combined modality therapy (CMT). A 21 years male patient presented with left sided facial swelling of 3 months duration along with proptosis, nasal intonation of voice and difficulty in opening the mouth. On examination, a single diffuse swelling noted on the left side of the face. CECT showed left sinonasal mass extending to nasopharynx, infratemporal fossa, left orbit and also to subcutaneous plane. He was managed by left sided maxillectomy followed by adjuvant chemotherapy and radiotherapy. EGGCT mostly occours in mediastinum followed by abdomen, mostly sacrococcygeal region. EGGCTs are most common in paediatric age group (two third of GCT is extragonadal), adults and adolescents mostly suffer from gonadal GCT. In this era of combined modality management surgery combined with chemotherapy and radiotherapy gives the best outcome.

Keywords: *Combined modality management, extragonadal germ cell tumor, germ cell tumor, maxilla*

Introduction

Germ cell tumors (GCTs) are neoplasms derived from germinal cells. Extragonadal GCTs (EGGCTs) are that which takes origin outside the gonads, which may be due to birth defects, resulting from errors during development.[1] There are controversies regarding the origin of EGGCTs.[2] Unlike adult and adolescents, GCTs in children are relatively uncommon, presenting approximately in 3% of all pediatric malignancies.[3] EGGCTs constitute only 1%–5% of all gonadal GCTs and EGGCTs.[4] Mediastinum is the most common anatomical site for EGGCTs in adults,[4] and it becomes very rare when it arises from the maxilla. These patients of maxillary GCT typically presented with facial swelling, nasal blockade, proptosis nasal intonation of voice, difficulty in breathing, difficulty in opening, mouth, etc. Maxillary GCTs commonly occur within the first three decades of life and males are more prone to develop this. When treated with neoadjuvant chemotherapy, with or without adjuvant surgery or radiation, patients with pure seminomatous EGGCTs have a long-term cure rate of almost 90%, irrespective of the primary tumor site. Patients with mediastinal nonseminomatous variety have 45% 5-year survival rate, whereas patients with retroperitoneal primaries have a better 5-year survival rate that is 62%.[5]

Case Report

A 21-year-old male patient presented at our institution with swelling of the left-sided facial swelling of 3-month duration followed by development of proptosis, nasal intonation of voice, and difficulty in opening the mouth. On further asking, he complained of blurring of vision of the left eye of 1-month duration. He visited otorhinolaryngologists outside in the past and was advised antibiotics but of no relief and the swelling increased with time.

On examination, a single diffuse swelling was present on the left side of the face, measuring approximately 4 cm × 3 cm in dimensions. The swelling was firm, tender, smooth, and immobile and was fixed to the underlying bone. On intraoral examination, a solitary diffuse swelling was seen on palate extending

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from the left first molar to the third molar and medially almost to midline. The surface of the swelling was smooth, reddish with no ulcerations. No clinically palpable lymph node was seen.

Contrast-enhanced computed tomography (CECT) showed large enhancing left sinonasal mass extending posteriorly to nasopharynx, laterally to infratemporal fossa and superiorly to upper limit of the left orbit. Anteriorly, the lesion also extends to the subcutaneous plane. Extensive bone erosion is seen. Bilateral neck levels IB, II, and V lymph nodes are also enlarged [Figure 1].

Histopathology [Figure 2] shows germ cell component of primitive neuroendocrine origin infiltrated all over; it also shows the glands as well as stroma infiltrated with plenty of germ cell component suggestive of malignant teratocarcinoma.

Management was done by the left-sided maxillectomy followed by adjuvant chemotherapy and radiotherapy. A conformal radiotherapy was planned and given, keeping in mind all the dose constraints of the organ at risk, which is shown in Figures 3 and 4.

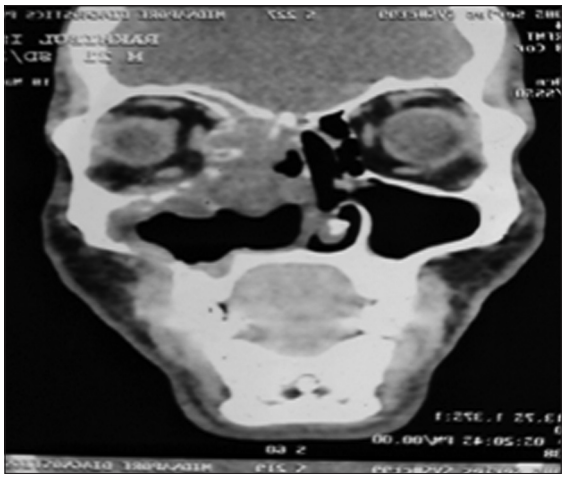

Figure 1: Contrast-enhanced computed tomography shows large enhancing left sinonasal mass extending posteriorly to nasopharynx, laterally to infratemporal fossa, and superiorly to upper limit of left orbit. Anteriorly, the lesion also extends to subcutaneous plane. Extensive bone erosion is seen. Bilateral neck node levels IB, II, and V lymph nodes are also enlarged

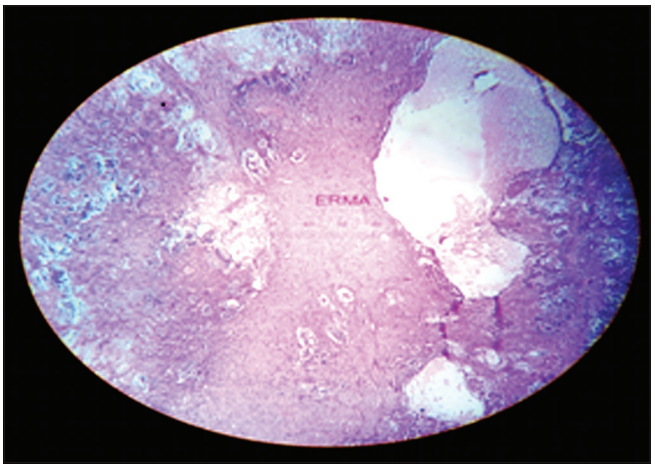

Figure 2: Histopathology shows germ cell component of primitive neuroendocrine origin infiltrated all over. This image shows the glands as well as stroma infiltrated with plenty of germ cell component suggestive of malignant teratocarcinoma

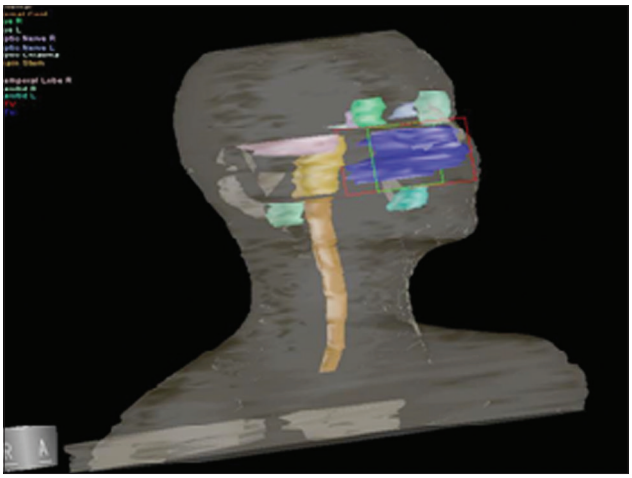

Figure 3: Three-dimensional reconstructed image for conformal radiotherapy planning

Discussion

As GCT of maxilla is a rare case, opportunity of case reporting and its management is of great importance. EGGCTs mostly occur in mediastinum followed by abdomen (mostly sacrococcygeal region). EGGCTs are most common in the pediatric age group (two-thirds of GCT is extragonadal), whereas adults and adolescents mostly suffer from gonadal GCT.[6] GCT at head–neck region is extremely rare, making it only 6% of all GCTs.[7]

Controversy remains regarding the origin of EGGCTs. EGGCTs are generally found in the midline of the body, e.g., retroperitoneal, anterior mediastinum, sacrococcygeal area, and the pineal gland. Less common sites are orbit, suprasellar area, palate, thyroid, submandibular and maxilla region, anterior abdominal wall, stomach, liver, vagina, and prostate. The classic pathophysiological theory till date is transformation of misplaced primordial germ cells during embryogenesis.[8]

In this modern era of combined modality management, surgery combined with chemotherapy and radiotherapy gives the promising outcome. It has been seen that chemotherapy increases the disease-free survival in testicular GCT.[9] This also implies for the management of EGGCTs. Results from international analysis showed that there is role of radiotherapy in postoperative setting where there is positive margin and/or residual disease.[10] However, till date, surgery whenever possible is the treatment of choice for EGGCTs and gives a better result.[11]

Conclusion

EGGCT of maxilla is a rare case. Diagnosis is done by CECT and histopathology. Surgery remains the mainstay of

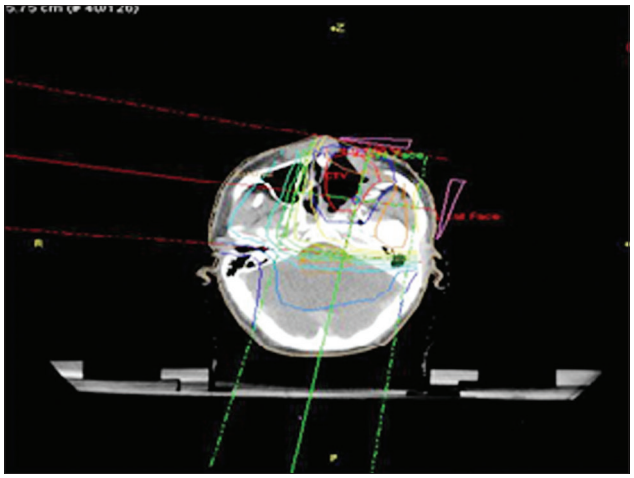

Figure 4: Three-dimensional conformal radiotherapy planning and beam arrangement

management of EGGCT. High risk features in postoperative histopathology guide for selecting adjuvant chemotherapy and postoperative radiotherapy. Adjuvant chemotherapy and radiotherapy after surgical resection decrease the chance of disease recurrence, though clear-cut guidelines are not yet available.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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