Case Report

Anterior mediastinal mass with superior vena cava syndrome: A rare presentation of germ cell tumor

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Abstract

The objective of this case report is to highlight the clinical, radiological, and histopathological features of a case of a 33-year-old male patient, who presented to AVBRH, Sawangi (Meghe), Wardha, with an anterior mediastinal mass with superior vena cava syndrome and after detailed studies was diagnosed as a case of germ cell tumor which was further confirmed on immunohistochemistry staining.

Keywords: Alpha-fetoprotein, anterior mediastinal mass, germ cell tumor, immunohistochemistry, superior vena cava syndrome

Introduction

Germ cell tumors of the mediastinum are a class that includes malignant and benign tumors that have their origins from the primitive germ cells that persist in the mediastinal region during the early developmental phase. The most prevalent extragonadal initial location for germ cell malignancies is the anterior mediastinum.[1]

Primary gonadal and mediastinal GCTs include histological and biochemical hallmarks, as well as cytogenetic abnormalities on isochromosome 12p.[1] Mediastinal germ cell tumors (MGCTs) are unique in that they are linked to Klinefelter syndrome and hematologic malignancies.[2] Teratomas, seminomas, and non-seminomatous tumors are the histological classifications.[3] Each categorization has significant differences in terms of prognosis and therapy.

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The majority of MGCTs are discovered by chance during imaging examinations. About 20% to 40% of the cases are completely symptom-free when they are first diagnosed. The expansion of the mediastinum and its protrusion into adjacent tissues are usually the first indications. Clinical indicators include cough, chest pain, breathlessness, nocturnal sweats fever, and weight loss. The symptoms are determined by the tumor's histological subtype and size. Depending on the histological subtype, initial surgical resection or cisplatin-based chemotherapy followed by surgical resection are the therapeutic strategies.

Germ cell malignancies are most usually seen in the reproductive organs,[4] primarily in the testicles, and in the ovaries only rarely. GCTs can be discovered outside of the gonads in midline regions such the mediastinum and retroperitoneum, as well as less frequently in the suprasellar and pineal locations. The source of germ cell cancer outside the realm of the reproductive organs remains unclear, despite a number of theories. During embryonic development, germ cells that are classified as primordial must migrate to the dorsally located urogenital ridge from the proximal epiblast for the initiation of testicular creation.[5] According to the dominant theory, they come to a

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halt during their descent, which then leads to the creation of a tumor in the mediastinum.[6]

An alternative hypothesis is that altered germ cells in the reproductive organs migrate backwards.[7] Although common genetic cell origin evidence supports this concept, it does not explain for some of the reported biological inconsistencies.

Case History

A 33-year-old male came to casualty on 30/8/21 with the chief complaints of dry cough, back ache, chest pain, anterior chest wall swelling, breathlessness MMRC grade I-II, dysphagia, and hoarseness of voice since 3 months.

On examination, the patient was normothermic, with a heart rate of 90/min, a respiratory rate of 20 cycles/min, a B.P of 136/90 mm Hg, and a Spo2 of 96% without oxygen. On systemic examination in respiratory system: bilaterally decreased breath sounds with occasional wheeze. No significant findings in cardiovascular, central nervous system and per abdominal examination.

Chest X ray PA view was done [Figure 1], which was suggestive of a large anterior mediastinal mass.

CECT Thorax was done and the findings were as follows

Large lobulated soft tissue mass lesion measuring 18 × 8 cm in axial dimension and 16 cm in craniocaudal extent seen centered within the superior and anterior mediastinum compressing trachea with moderate luminal narrowing, 2.2 × 1.1 cm sized fat-attenuation area with small calcification noted within the lesion, contiguous extension of the lesion into the anterior chest wall on the right side, suspicious of neoplastic etiology [Figures 2 and 3].

Patient had signs of SVC obstruction. His lab investigations were as follows: Hemoglobin: 11.9 gm/dL (Normal range - For men, 13.2 to 16.6 grams per deciliter), WBC count: 3900 × 10^9/L (Normal range - 4,500 to 11,000 WBCs per microliter (4.5 to 11.0 × 10^9/L), Platelets 1,54,000/microliter (Normal range - 150,000 to 450,000 platelets per microliter of blood), Tumor biomarkers- Serum Alpha-fetoprotein (AFP) levels were 3.66 ng/mL (Normal range- between 10 ng/mL and 20 ng/mL is normal for adults), beta-HCG levels were raised - 167.08 (Normal range - <5 IU/L). Computed tomography (CT)-guided biopsy [Figure 4] was done and histopathological examination showed sheets of severely atypical neoplastic cells (malignant) intermixed with areas of hemorrhage and necrosis will cells being suspicious of germ cell origin on histopathology and immunohistochemistry for confirmation of diagnosis and management. Immunohistochemistry staining was done of the biopsy sample and cells were found to be immunoreactive for Oct-3/4 and negative for CD-45 and the diagnosis of germ cell tumor was confirmed. The patient's SVC syndrome showed considerable resolution within two weeks of chemotherapy and radiotherapy.

Discussion

To examine a patient suspected of having MGCT, a complete physical examination is required, with specific focus paid to the gonads. To detect co-existing GCTs, routine screening should include testicular ultrasound. Advanced biochemical, radiological, and histological studies[3] are of utmost importance to confirm the diagnosis and to provide prognostic accuracy. Patients frequently arrive after a chest X-ray reveals an incidental anterior mediastinal tumor. CT or magnetic resonance imaging (MRI) can be used to determine the tumor's borders, dimensions, and the exact location.[8] The biochemical parameters that may be raised and that require special diagnostic considerations are: Lactate dehydrogenase (LDH), beta-human chorionic gonadotropin (b-hCG), and alpha-fetoprotein (AFP) levels. The therapy for MGCTs is identical to the treatment for gonadal GCTs. Because of the disease's rarity, large prospective

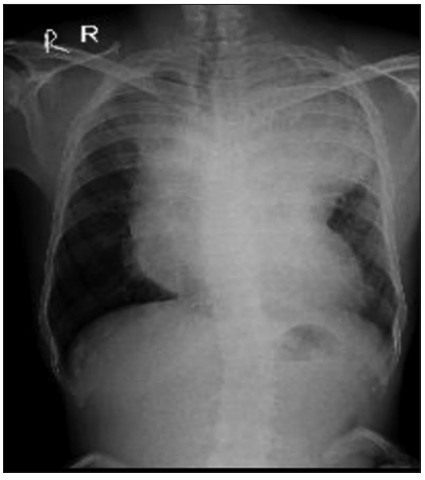

Figure 1: Chest X-ray PA view showing a large anterior mediastinal mass

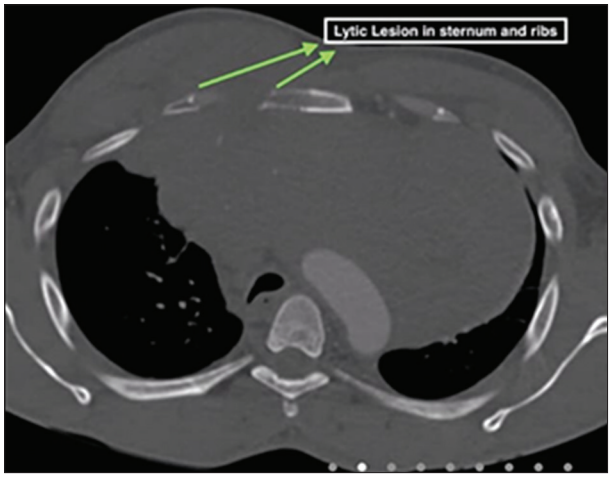

Figure 2: CECT Thorax - large lobulated soft tissue mass lesion in superior and anterior mediastinum with lytic lesions in the sternum and ribs

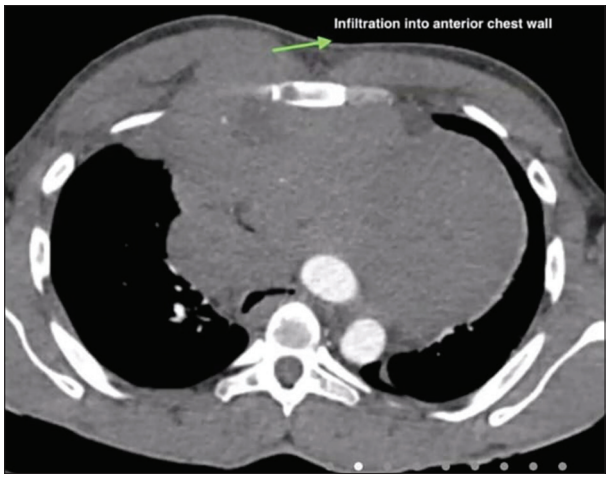

Figure 3: CECT Thorax - large lobulated soft tissue mass lesion in superior and anterior mediastinum with infiltration into the anterior chest wall

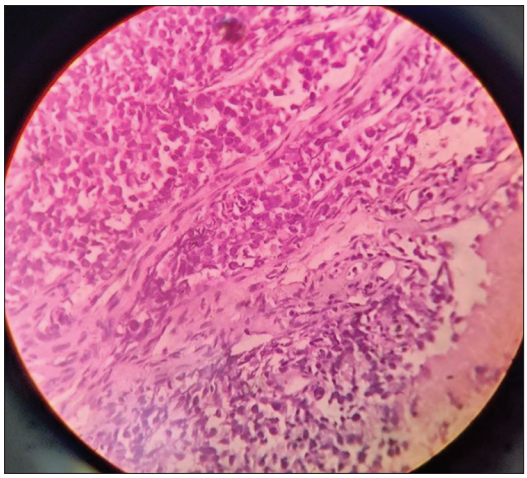

Figure 4: CT guided biopsy from the mediastinal mass suggestive of severely atypical malignant cells with areas of hemorrhage and necrosis

randomized controlled trials have been difficult to conduct. The International Germ Cell Cancer Collaborative Group classifies non-seminoma MGCTs as "low risk" tumors (IGCCCG),[9] while the Seminoma and Teratomas MGCT are sub-classified as excellent or intermediate risk based on the definitive evidence of metastasis and tumor markers.[9] When benign mature teratomas[10] become symptomatic, surgical intervention becomes necessary since they do not respond to medical management.[11] A posterolateral thoracotomy or a median sternotomy are the main approaches used in surgical resection. Because these tumors seldom exhibit extra-mediastinal invasion, incomplete resection to minimize essential structural involvement does not necessitate further chemoradiotherapy. It is important to recognize the early manifestations of SVC syndrome and a mediastinal mass when they first present to a primary practitioner so as to arouse a high index of suspicion for early referral to a specialist for optimal management, especially given the tendency of a SVC syndrome patient to deteriorate if rapid therapeutic measures are not implemented.

Differential Diagnosis

- Thymic carcinoma[12]
- Sarcoma
- Thymoma
- Lymphoma
- Myasthenia gravis
- Tuberculosis

Conclusion

Age, the number of sites where metastasis has occurred, tumor elevation, and biochemical parameters are all prognostication factors (LDH, AFP, b-hCG). Elevated AFP levels and raised b-hCG levels at the time of diagnosis are associated to even worse outcomes. Patients with seminoma GCTs had a 72% to 100% five-year survival rate, compared to 48% to 65% for patients with non-seminomatous GCTs. For non-seminomatous malignancy, young patients (under 29-year-olds) with normal b-hCG levels had the best outcomes. Non-pulmonary metastasis in the visceral organs is associated with a poor prognosis.[12] Seminomatous tumors without involvement of the lungs were associated with a five-year survival rate upwards of 90%. Future studies regarding the newer diagnostic modalities for the diagnosis of germ cell tumors are eagerly awaited.

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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