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Case Report

Mediastinal Seminoma presenting as superior vena cava syndrome and tracheal obstruction

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Abstract

Malignant germ cell tumours of the mediastinum are rare, presenting mostly in young males. These are bulky tumours, mostly intrathoracic, infiltrating into adjacent structures early in the growth process. Patients may present with symptoms of compression. Occasionally, mediastinal adenopathy and superior vena cava (SVC) syndrome may occur.

We discuss a case of a 19 year old boy with six weeks history of progressively worsening shortness of breath and mid-sternal chest heaviness and one month history of swelling of the face and upper limbs. The entire face and upper extremities were grossly oedematous with engorged veins suggesting SVC syndrome. CT scan chest showed a large mediastinal mass 18x24 cm extending from the superior mediastinum to the mid pericardial area. The trachea was extrinsically compressed to almost 80% at its distal portion. Mass was resected completely. Post operative course was unremarkable. The final histopathology showed a fibrotic mass with no viable tumour. Surgery followed by radiation therapy has been the usual initial treatment with multimodal chemotherapy showing remarkable results.

Introduction

Primary malignant germ cell tumours of the mediastinum are rare.^{1,2} Only 2-5% of germ cell tumours occur at extra-gonadal sites.³ Seminomas are malignant germ cell tumours that account for about 25% of all primary mediastinal germ cell tumours.⁴ They are speculated to arise in unusual locations due to migration of germ cells during embryogenesis.⁵ These tumours are more common in young males usually occurring in the third or fourth decade of life.²

Seminomas are radiosensitive tumours, yet radiotherapy is associated with a higher risk of recurrence. The introduction of Cisplatin based chemotherapy, as initial treatment has greatly improved the prognosis of these tumours.

Case Report

We present the case of a 19 year old boy presenting to the ER, with six weeks history of progressively worsening shortness of breath and mid-sternal chest heaviness and one

month history of swelling of the face and upper limbs. On examination, he was in extreme distress with tachycardia and tachypnoea. He had an Oxygen saturation of 88% on room air. The entire face and upper extremities were grossly oedematous with engorged veins suggesting SVC syndrome. Patient had a CT scan chest done, a week prior to this presentation which showed a large mediastinal mass, 18 cm by 24 cm, extending from the superior mediastinum to the mid pericardial area. It was completely occluding the superior vena cava and innominate vein. The trachea was extrinsically compressed to almost 80% at its distal portion. An urgent biopsy of this mass was done without anaesthesia with BIPAP (Bi level positive airway pressure) support in the OR and alpha fetoprotein, beta HCG and LDH levels were sent. Within 24 hours the biopsy results revealed mediastinal seminoma. Patient was started on BEP (Bleomycin Etoposide Cisplatin) regimen chemotherapy and within 48 hours patient became better and was able to be discharged without any oxygen after one week, with almost complete resolution of his SVC syndrome and tachypnoea. Patient received six cycles of chemotherapy after which via median sternotomy the residual mass was resected completely. Post operative course was unremarkable. The final histopathology showed a fibrotic mass with no viable tumour.

Discussion

Mediastinum is the most common site for extra gonadal germ cell tumours, which represent about 10-15% of mediastinal tumours.³ Other extra-gonadal sites that can be involved are the retroperitoneum and the pineal gland.⁵ The exact pathogenesis of germ cell tumours in the mediastinum is unclear. Several theories exist of which two have gained wider acceptance.² The germ cell theory proposed by Friedman suggested that these tumours arise from germ cells that migrated to the thymus during embryogenesis. He also proposed that germ cells may be distributed to the thymus, brain, liver and bone marrow. Schlumberger hypothesized that the tumours arise due to an abnormality in thymic development during embryogenesis.^{1,2} More recently Rosai et al suggested that they may arise from myoid cells present in the thymus.¹

Seminomas are slow growing, bulky tumours that invade locally early in the growth process. Patients may

present with symptoms of compression of surrounding structures, like dyspnoea, dysphagia, cough and chest pain, or simply constitutional symptoms like fever and weight loss. Uncommonly, mediastinal adenopathy and superior vena cava syndrome may occur.² About 20-30% of patients are asymptomatic.^{1,2,6} Teratoma, primary large B cell lymphoma and thymoma are other differential diagnosis.⁴

The most common sites of metastases of the tumour are the lungs and regional lymph nodes. Other sites that have been reported are the skeletal system, tonsils, thyroid gland, spleen, skin, brain and spinal cord.² Metastasis of testicular seminomas to the mediastinum is rare and has been documented in only a few cases.

Extragenital germ cell tumours are histologically similar to, but differ biologically and in clinical outcome from their gonadal counterparts.^{3,7} Some distinguishing histologic features, however, like remnants of thymic tissue and cystic changes similar to multi-locular thymic cysts may be identified in mediastinal seminomas.¹ Diagnosis is by a combination of radiographs, computed scans and tumour markers (elevated alpha fetoprotein and beta HCG are commonly found), and biopsy for histologic confirmation. The overall survival rate for mediastinal seminomas is about 70-75%.^{1,2}

Since a long time, radiotherapy with or without surgery has been considered the primary modality of treatment for seminomas because of their radiosensitive characteristics.³ However this has been associated with a high rate of recurrence with about one third of the patients relapsing.⁸ Since both surgery and radiotherapy aim at achieving local control, recently chemotherapy has been advocated as the primary modality in treatment of mediastinal seminomas. Some studies report long term disease free intervals in about 85% of the patients receiving chemotherapy, compared to 65% receiving radiotherapy.⁶

Before the availability of Cisplatin, the combination chemotherapy of bleomycin and vinblastine was effective in patients with non-seminomatous germ cell tumours but not in seminomas.⁷ Given the rarity of primary mediastinal seminomas, most studies are retrospective and contain cases of both seminomatous and non seminomatous tumours. However, they do show the effectiveness of chemotherapy in seminomas after the introduction of Cisplatin.⁸

Reported relapse rate of germ cell tumours with Cisplatin based chemotherapy is approximately 6%.⁹

Cisplatin based chemotherapy along with Etoposide and bleomycin is reported to have a cure rate of 90% in low risk patients and 70-80% in advanced seminoma, according to the International Germ Cell Cancer Collaborative Group Consensus Classification (IGCCCG). Prognosis is less clear for patients in the intermediate risk group with liver and CNS metastasis.¹⁰

In our patient BEP (Bleomycin, Etoposide, Cisplatin) regimen chemotherapy proved successful with almost complete resolution of his symptoms within 48 hours. Residual mass was resected to assess response to chemotherapy and to remove chemotherapy resistant disease. Histopathology of the resected residual mass also did not show any viable tumour. The patient was disease free until last follow up.

Our case shows the successful outcome of using Cisplatin based chemotherapy followed by debulking surgery to treat primary mediastinal seminoma.

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