NON SEMINOMATOUS GERM CELL YOLK SAC TUMOR OF THE ANTERIOR MEDIASTINUM

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SUMMARY: A case of yolk sac tumor of the anterior mediastinum, which presented with superior vena cava (SVC) syndrome and associated respiratory manifestations, is presented. The steps of diagnostic procedures, surgical treatment, as well as post-operative management, were discussed within the scope of the current literature.

Key Words: Mediastinal Masses, Yolk Sac Tumor.

INTRODUCTION

Gerin cell tumors most often occur in the gonads but may occasionally arise in other areas, such as the mediastinum. The extra gonadal cell tumors are thought to arise from primordial gerin cells of the nephrogenic ridge (1).

Histopathological classifications of germ cell tumors include seminoma, embryonic carcinoma, teratocarcinoma and endodermal sinus or yolk sac tumors (2). The rarest of these tumors mentioned above is endodermal sinus yolk sac - tumor. Elevated levels of either \(\beta\)-Human chorionic gonadotropin (\(\beta\)-HCG) or Alpha-fetoprotein (AFP), or both are usually found in these patients.

Non-seminomatous malignant germ cell tumors are less common than seminomas. They are rarely diagnosed in the anterior mediastinum.

In this paper, we present a case of yolk sac tumor of the anterior mediastinum with clinical presentation of the symptoms of superior vena cava syndrome as well as respiratory problems.

CASE REPORT

A 36-year-old male was admitted to a foreign hospital, with the complaints of dyspnea on exertion, hemoptysis, nocturnal sweating and loss of weight for the previous 2 months.

On admission, physical examination revealed signs of SVC syndrome and a reduction of respiratory sounds in the right hemithorax. Routine laboratory data were within normal limits except white blood cell count (12.500 / ml) and erythrocyte sedimentation rate (122 mm / h).

Chest X-ray, thoracic computerized tomography (CT) and magnetic resonance imaging (MRI) revealed a heterogenous anterior mediastinal mass $13.5 \times 14 \times 12$ cm in size. This mediastinal mass caused compression on the ascending aorta, SVC and neighboring pericardium as well as the right hilum (Fig. 1A, B, C).

Bronchoscopic lavage demonstrated malignant cells and a CT guided fine needle biopsy was performed. Histopathology revealed poorly differentiated squamous cell carcinoma.

As a result the patient underwent radiotherapy with a dose of 4000 - 5000 rads.

The patient was then referred to our hospital for further examination and treatment.

On admission to our hospital, the symptoms were still present and the mass was observed to diminish on reevaluation by CT (13x14x10 cm.). The AFP level was reported to be 300 IU/ ml. Routine laboratory examinations were very similar to the previous ones.

Considering the results of the histopathology and radiology, the patient was scheduled for operation.

Operative procedure:

After premedication with diazepam (10 mg I.M.), a radial artery catheter, two peripheral intravenous catheters and a subclavian vein catheter were inserted in the operating room.

Hemodynamic parameters; heart rate, mean arterial pressure, central venous pressure and arterial blood gases were monitored throughout the procedure.

Anesthesia was induced by fentanyl (15 mcg/kg) and muscle relaxation was established with pancronium (0.1 mg/kg). The patient was were intubated endotracheally and ventilated with 100% oxygen.

Following right thoracotomy via 5th intercostal space; a huge, round, capsulated mass was observed in the anterior mediastinum compressing the SVC, right lung, ascending aorta and encroaching on the pericardium. The mass was totally removed from the surrounding structures along with a part from the involved pericardium. The specimen was reserved for histopathological examination.

The operation was then completed by standard procedure.

RESULTS

The early postoperative completed by of the patient was uneventful.

Histopathology and immunohistochemical

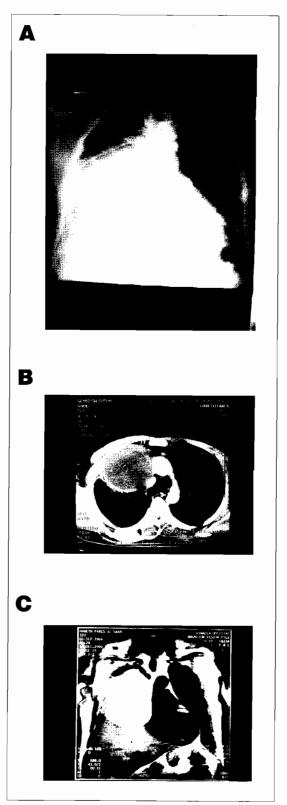


Fig. 1: Chest X-ray showing right opacity (A). Computerized tomography of the chest showing anterior mediastinal mass (B). MRI of the thorax showing anterior mediastinal mass (C).

evaluation confirmed an infantile yolk sac tumor with negative HCG and strong positive AFP results (Fig. 2). The AFP level which was 300 IU/ ml pre-operatively declined to 62.9 IU / ml post-operatively.

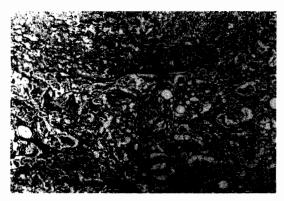


Fig. 2: Yolk sac tumor: Atypical elements with soft reticular pattern and papillary structures.

Postoperative chemotherapy included Etopsoid, Cisplatin and Bleomycin at standard doses.

The late postoperative follow-up was uneventful and the patient is free of symptoms 8 months after the operation.

DISCUSSION

Primary mediastinal tumors and cysts are common in young and middle-aged patients. Most masses are discovered on routine radiographic examinations in symptomatic patients, but many lesions produce non-specific clinical manifestations.

Endodermal sinus (yolk sac) tumors are commonly located in the anterior mediastinum producing the compression symptoms. The diagnostic procedures depend on the location and the tumor markers that are secreted from the relevant type of the tumor. The anterior mediastinal masses associated with high levels of AFP and HCG should arouse the suspicion of possibility of a germ cell tumor. In our case we also followed the same procedure.

Although germ cell tumors are highly radiosensitive and are sometimes cured solely with radiotherapy, results using this method alone have been disappointing because these tumors frequently are very large, or because of evidence of metastatic spread at the time of diagnosis, or both (3). Several investigators have reported improved results in patients who underwent surgical resection followed by radiotherapy (4).

The combination therapy also has markedly altered the approach to the treatment of non-seminomatous germinal tumors in recent years. The combined use of extensive surgical removal and chemotherapy has been demonstrated to be curative in many patients with non-seminomatous mediastinal germinal tumors. Extra gonadal non-seminomatous germinal neoplasms, while formerly fatal in most instances, can now often be cured using combined chemotherapy with aggressive surgical resection (5).

We have also discussed the indications of the operation and concluded that this patient might benefit from surgery. Postoperative follow-up also confirmed our decision.

However, we believe that when initial resection is impossible or unsafe, combination chemotherapy should be the initial treatment, followed by reexploration if response to therapy is incomplete.

Our patient underwent radiotherapy postoperatively in order to have a conservative debulking and a safe surgical result. Combined chemotherapy was employed after the operation with a cisplatin containing regimen as expressed in the literature.

This combined therapeutic policy in yolk sac tumor cases has been a recent approach, and the outcome is still controversial since there are no studies with a high number of cases. However we think this kind of treatment would at least provide the stability in the prognosis as in our patient.

Further complementary studies with a large number of cases may illuminate the appropriate protocol. Correspondence to: Velit HALİT, M.D.

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