Case 13108

Primary posterior mediastinal yolk sac tumour in a two-year-old girl
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Section: Paediatric radiology
Area of Interest: Thorax Oncology Paediatric
Procedure: Decision analysis
Procedure: Diagnostic procedure
Technique: Digital radiography
Technique: Ultrasound
Technique: CT
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 2 years, female

Clinical History:
A 2-year-old girl was referred to our department for respiratory distress. There was no relevant information in her past medical history. Physical examination revealed respiratory distress with intercostal and sub-ternal retractions, and tachycardia. The right hemithorax examination showed decreased movements, absolute dullness and suppressed breath sound. The AFP level was remarkably high.

Imaging Findings:
Chest X-rays showed a right opaque hemithorax with a mediastinal shift due to a large right pleural effusion (Fig. 1). A chest and lung ultrasonography (US) was performed and showed a hyperechoic mass with dented edge located in the mid inferior part of the right hemithorax associated with a pleural effusion [2]. The patient was transferred to cardiovascular surgery department because of the worsening of her respiratory state. An exploratory thoracotomy was urgently performed and revealed a fragile mediastinal mass invading the parietal pleura. Resection of the tumour was not attempted. Instead, biopsies were taken and a drain tube was left. CT-scan (computed tomography) revealed a large posterior mediastinal mass spreading in the inframediastinum posterior space, repressing cardiac cavities and invading the adjacent pleura and the diaphragm (Fig. 3 a, b, c, d and Fig. 4). A capsular rupture was noticed. Mediastinal and coeliac lymphadenopathies were detected in addition to pleural and hepatic metastasis.

Discussion:
Yolk sac tumour (YST) is a primitive malignant germ cell tumour (GCTs) that is uncommon in clinical practice and seems to have a special predilection for males [1]. It occurs mostly between one to five years of age involving most frequently gonads than extragonadal sites [2] with a predilection of anterior mediastinum [3]. Our report contradicts common literature data as far as our patient is female and the tumour is localized at the posterior mediastinum. To our knowledge, this is the second female patient case with a primary posterior mediastinal YST. There are no specific clinical signs for YST.
The finding of an elevated AFP serum level is useful in the diagnosis and follow-up of these patients [3] and should be measured at presentation and monitored during treatment. They are very useful in diagnosis and in evaluating the effectiveness of therapy [4]. The dosage of this parameter can be sufficient to the diagnosis without histological
confirmation [4]. The rate should be superior to 1000 kU/L. Radiology can aid in surgical planning, and avoid inappropriate management, and may enable the surgeon to plan the adequate procedures [1].

According to Li and Al [1], the CT (Computed Tomography) aspect of YST ranges from entirely solid to predominantly cystic or presents as heterogeneous appearances consisting of a mixed solid and cystic nature and tend to grow in an expansive centripetal fashion with compression of adjacent organs. On unenhanced CT, the solid portion of YST appeared as isodense to normal muscle. Enhancement can be more or less marked. Its heterogeneity may be due to intraloesional haemorrhage, necrosis, or cystic change. CT can also pinpoint capsular rupture which may lead to a large spread into the adjacent structures, such as abdominopelvic cavity, surrounding soft tissue space, pleural cavity, lung parenchyma, or tracheobronchial tree. This may cause inflammation, adhesion, metastasis implantation, or invasion and compression of adjacent organs. Intratumoral calcification and fatty tissue, although rare, may indicate a mixed YST containing a teratoma component.

YST are chemosensitive. Surgical excision is also an important step for the treatment of these tumours [4]. Radiotherapy has no place in YST management [4]. Surgery increased the response rate to 42% [4]. Analysis of the operative specimen suggests that surgical removal is necessary after chemotherapy as far as follow up cannot be based solely on AFP rate. Prognosis is intimately linked to the rapid onset of the treatment and to the initial extension of the tumour.

**Differential Diagnosis List:** Yolk sac tumour arising within the posterior mediastinum, Rhabdomyosarcoma, Teratoma

**Final Diagnosis:** Yolk sac tumour arising within the posterior mediastinum

**References:**


Figure 1

Description: Near-complete opacification of the right hemithorax with pleural fluid. Note displacement of left paraspinal stripe (arrow). Mild shift of heart to left side is seen (star). No rib erosion identified. Origin: Douira Khomsi W, Departement of Radiology, Bechir Hamza infant hospital, Tunis, Tunisia.
**Figure 2**

Description: heterogeneous echogenic mass (arrow) in the right pleural cavity surrounded by pleural fluid (star). **Origin:** Douira Khomsi W, Departement of Radiology, Bechir Hamza Infant Hospital, Tunis, Tunisie.
Description: Heterogeneous mass with the extension into the pleural cavity and inferiorly into the liver. Aerated lung and small amount of collapsed lung are at the superior aspect of the mass. Origin: Ammar I, Departement of Radiology, Bechir Hamza Infant Hospital, Tunis, Tunisie.
Figure 4

Description: Follow-up radiograph after 1 course of combination chemotherapy. Origin: Fedhila F, Departement of peadiatrics, Bechir Hamza Infant Hospital, Tunis, Tunisie.
Figure 5

**Description:** Large pleural effusion (star), repulsing the right lung. **Origin:** Douira, W, Departement of Radiology, Bechir hamza infant's hospital, Tunis, Tunisia

**Description:** Heterogeneous attenuation mass causing the shift of the mediastinum. **Origin:** Douira, W, Departement of Radiology, Bechir hamza infant's hospital, Tunis, Tunisia
**Description:** Heterogeneous attenuation mass causing the left shift of the heart (arrow). **Origin:** Douira, W, Departement of Radiology, Bechir hamza infant's hospital, Tunis, Tunisia

**Description:** Axial contrast-enhanced CT images at the lung bases reveal the heterogeneous mass (M) in the hepatic hilum consistent with hepatic invasion (arrow). **Origin:** Douira, W, Departement of Radiology, Bechir hamza infant's hospital, Tunis, Tunisia