CASE REPORT

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Unusual finding of a mediastinal T-cell lymphoma in a 13-year-old patient – a case report

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Abstract

T-cell lymphoblastic lymphoma is an aggressive malignancy that represents 85% of all lymphoblastic lymphomas. It usually occurs in late childhood, adolescence and young adulthood with a 2:1 male preponderance and it presents with pleural effusion and respiratory symptoms and in rare cases vena cava syndrome can be encountered. We present the case of a 13-year-old patient who was referred to our clinic from a local hospital where he was diagnosed with a mediastinal tumor. The patient presented with thoracic pain, fever, coughing and fatigability for a month prior to admission, after having underwent surgery for abdominal pain (appendectomy). On admission to our hospital, a thoracic computed tomography (CT) scan was performed and showed the presence of an anterior mediastinal mass measuring 109/76/140 mm, well defined, which came in close contact with the superior vena cava, the ascending aorta and the pulmonary artery, right pleural effusion and a collapsed lung on the right side. The decision was taken to perform a tumor biopsy and a right pleural drain was placed. The patient's post-operative evolution was favorable with the remission of the respiratory symptoms. The histopathological result showed the presence of T-cell lymphoblastic lymphoma and the patient was then transferred to the oncology ward where he underwent chemotherapeutic treatment, with a favorable outcome. T-cell lymphoblastic lymphoma is an aggressive type of lymphoma and it is usually hard to diagnose considering the fact that the symptoms are often vague. It is essential to establish the diagnosis without delay and start appropriate chemotherapeutic treatment.

Keywords: lymphoblastic lymphoma, pleural effusion, mediastinal biopsy.

Introduction

T-cell lymphoblastic lymphoma (T-LBL) is an aggressive malignancy and represents 85–90% of all lymphoblastic lymphomas [1–3]. It usually occurs in late childhood [1], adolescence and young adulthood with a 2:1 male preponderance T-LBL frequently presents as a mediastinal mass, with secondary localizations such as skin, liver, spleen, central nervous system or testis, although these sites without mediastinal involvement is rare [2, 4]. While pleural effusion and mediastinal adenopathies are common signs of T-LBL, establishing an accurate diagnosis is often challenging. Treatment for T-LBL consists of chemotherapy and is usually divided into three phases: induction, consolidation and maintenance [2]. In children, the five-year survival rate with treatment is between 80–90% [3].

We present a case of T-LBL in a patient with a large mediastinal tumor, in whom clinical symptoms were unsophisticated for a long period of time.

Case report

We present the case of a 13-year-old patient who was referred to our clinic from a local hospital where he was diagnosed with a mediastinal tumor.

A month prior to admission the patient presented to a local hospital with fever and abdominal pain and he was diagnosed with acute appendicitis and underwent an emergency appendectomy. His post-operative evolution was favorable and three days after surgery he was discharged. A week later, the patient presented to the emergency room for thoracic pain, fever, coughing and fatigability and was admitted to hospital with the suspicion of pneumonia; a thoracic X-ray and a computed tomography (CT) scan were performed and showed pleural effusion and the presence of a mediastinal mass (Figures 1 and 2).

The patient was then referred to our clinic. On admission, the patient presented with an altered general state, fever (38.1^0C), 93% O_2 saturation, 40 respirations per minute, cough and an abolished vesicular murmur in the right hemithorax. A thoracic CT scan was performed and showed the presence of a well-defined, anterior mediastinal mass measuring 109/76/140 mm, which came in close contact with the superior vena cava, the ascending aorta and the pulmonary artery, massive right pleural effusion and a collapsed lung on the right side. An echocardiography was also performed and revealed pericardial effusion.

Under general anesthesia, an evacuatory pleural drainage

of the right thoracic cavity was performed and 300 mL of hematic fluid were drained. Microscopic examination of the fluid showed 80% lymphocytes present.

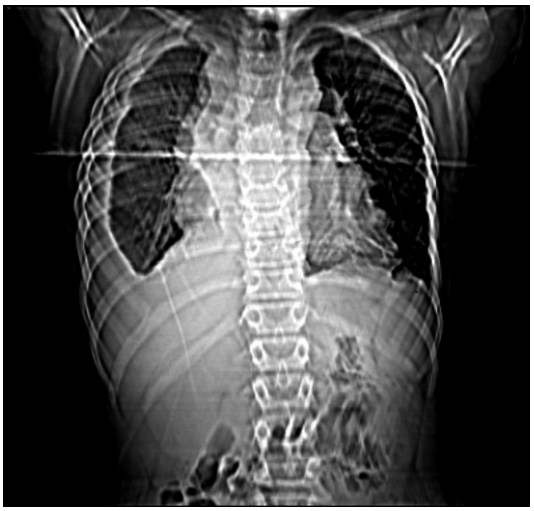

Figure 1 – ***Thoracic X-ray: mediastinal mass.***

Because the patient continued to present thoracic pain, respiratory distress and showed signs of superior vena cava syndrome, the decision was taken to perform a tumor biopsy and an internal mammary ganglia biopsy; a right pleural drain was also placed. The patient's post-operative evolution was slowly favorable with the remission of the respiratory symptoms.

The pathological results showed the presence of T-cell lymphoblastic lymphoma, with CD4-/CD8+ phenotype. Immunohistochemistry was positive for CD3, CD7, CD5, CD8 and CD1a, and negative for CD20, CD79a, CD4, CD10, TdT and CD99 (Figures 3–5). The patient was then transferred to the oncology ward. In order to evaluate the extension of the tumor, a bone marrow biopsy was performed which was negative. The patient was diagnosed with stage III T-LBL and he began the induction phase of the chemotherapeutic treatment, according to the Malignant Lymphoma Therapy (LMT) 96 protocol.

He has now begun the maintenance phase of the chemotherapeutic protocol and has had a favorable evolution, having shown no sign of relapse.

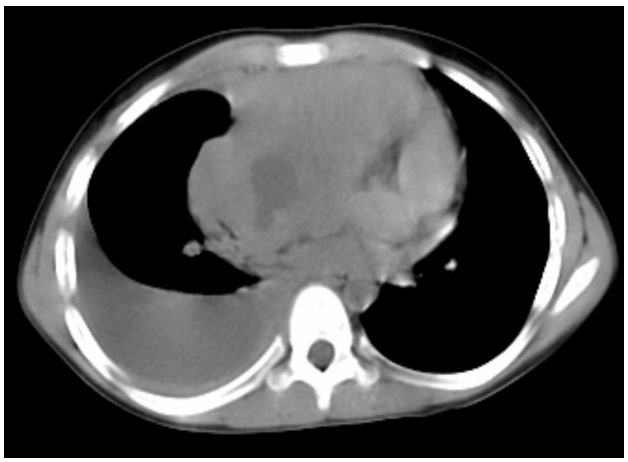

Figure 2 – ***CT scan: mediastinal mass with right pleural effusion.***

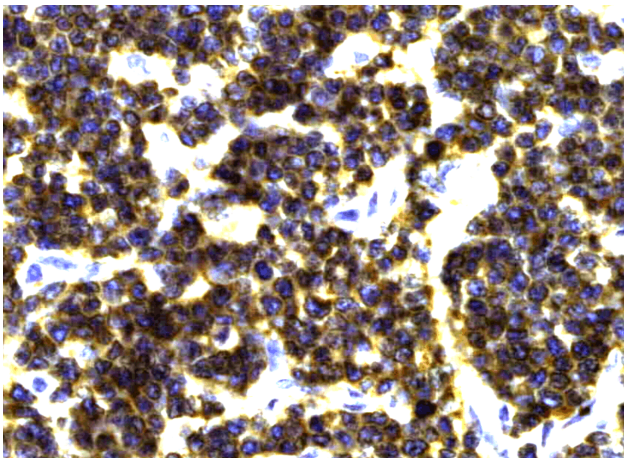

Figure 3 – ***Tumor cells with positive CD3. Immuno-marking with anti-CD3 antibody, ×200.***

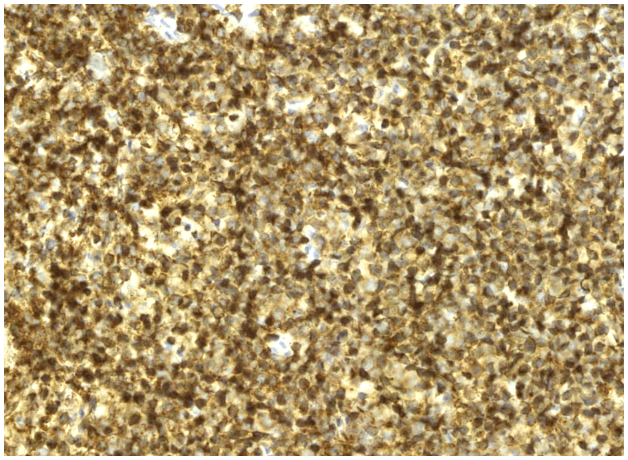

Figure 4 – ***Tumor cells with positive CD8. Immuno-marking with anti-CD8 antibody, ×100.***

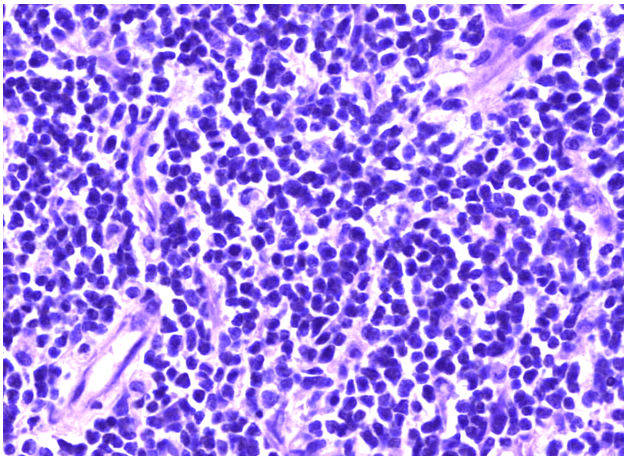

Figure 5 – ***Tumoral cells. HE staining, ×200.***

Discussion

Lymphoblastic lymphoma is a rare malignancy that represents less than 2% of all non-Hodgkin lymphomas, developing from T- or B-lymphoid progenitor cells [5]. T-LBL represents almost 85–90% of all lymphoblastic lymphomas and it is mostly encountered in male adolescents, only a small percentage of adult cases having been reported [1, 2]. These tumors occur most often in the anterior mediastinum and lymph nodes above the diaphragm [6, 7]. In 80% of cases, patients present with a mediastinal mass or bone marrow localization,

but secondary localizations such as the spleen, the liver, the testis or the central nervous system have also been encountered [2].

While some authors have reported cases of patients with unusual presentation of T-LBL such as a large breast mass or primary pleural lymphoma, most patient are diagnosed with an anterior mediastinal mass. They usually present to the emergency room with severe respiratory distress and nearly 70% also present with pleural effusion [8]. The respiratory symptoms in these patients are usually vague and can occur either because of the size of the effusion, the mediastinal mass that causes compression or both [9]. Nearly 60% of patients with pleural effusion often present with pericardial effusion [1].

Our patient is an adolescent male, who underwent emergency appendectomy for abdominal pain and after surgery, continued to present with recurrent subcostal pain, fever and fatigability. The challenge presented by our case was establishing a correct diagnosis. Taking into account the patient's medical history and his symptoms, the patient was admitted to hospital with the suspicion of pneumonia and the subsequent X-ray and CT scan showed the presence of a mediastinal mass and pleural effusion. According to a study published by Pietsch *et al.* [9], the differential diagnosis of malignant thoracic lymphoma can be made with pneumonia.

Because of the aggressiveness of this type of lymphoma, it is important to establish a correct diagnosis and begin appropriate treatment. Treatment for T-LBL consists of chemotherapy [3] and it is usually divided into three phases: induction, consolidation and maintenance [2], similar to those used in acute lymphoblastic leukemia. The prognostic of T-LBL is generally unfavorable in children; the five-year survival rate with treatment is between 80–90% [3], however, other studies have shown that the survival at five years should be much smaller. By other studies, the salvage rate for these patients is poor, with only a 14% overall survival [10]. Evolution does not seem to be influenced by factors such as age, gender, hepatomegaly, splenomegaly, associated infections, etc. However, the presence of pleural reaction to the accumulation of a large amount of pleural effusion and lymph nodes above the diaphragm are a low prognostic factor [11]. Pleurisy is a common complication of lymphoma [12].

Our patient presented right pleural effusion, 300 mL, hemorrhagic, in which we highlighted numerous lymphocytes that raised the suspicion of mediastinal lymphoma. Pleurisy has a multitude of pathogenic mechanisms. Most often mentioned are thoracic duct obstruction, airway obstruction mediastinal lymph drainage, lung infections, etc. [13, 14]. T-LBL relapses after treatment are common and have a dismal prognosis in both children and adults. Most patients suffer the first relapse after the second session of chemotherapy [15].

Conclusions

T-cell lymphoblastic lymphomas are aggressive lymphomas that are usually difficult to diagnose because the symptoms are often vague. Establishing a correct diagnosis and starting appropriate treatment is essential.

Conflict of interests

The authors declare that they have no conflict of interests.

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