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Case report

Askin tumor of thoracopulmonary region in a child: Case report and literature review

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ABSTRACT

Askin tumor is an uncommon malignant neoplasm of the thoracopulmonary region mainly occurring in children and adolescents. In this case, Computed Tomography (CT[1]) imaging features of Askin tumor and differential diagnosis criteria with others possible chest neoplasms are presented.

Introduction

CT with intravenous injection of contrast media represents an useful imaging technique for suspected neoplastic diagnosis and differential diagnosis of thoracic masses in children. Askin tumor is an uncommon malignant neoplasm in children and CT study can be used as a problem-solving tool.

Case report

A 7-year-old child with a history of persistent cough, weight loss and chest pain in the right side from about two months, admitted to our Pediatric Hospital. Physical examination revealed a decreased breath sound in the right lung. A chest radiography showed a large opacity occupying most of the right hemithorax with involvement of the lateral arch of the seventh rib that appeared with thinning of the bone matrix and sclerotic reaction of the cortical bone (Fig. 1). To better document this opacity, a CT with intravenous injection of contrast media was performed. CT images revealed a heterogeneous, voluminous and lobulated mass interesting the right seventh rib, which showed an inhomogeneous enhancement and necrotic and cystic degenerations within (Fig. 2A). A severe displacement of the mediastinal structures and infiltration of the diaphragm were demonstrated (Fig. 2B,C). CT bone windows improved the evaluation of the involvement of the lateral arch of the rib (Fig. 2D). A biopsy demonstrated the presence of the characteristic Homer Wright rosettes; a certain diagnosis of Askin tumor of the chest wall was made. The patient received a cycle of induction chemotherapy. After that, a CT scan was repeated. The axial plane and the Multi-Planar Reconstructions (MPR[2]) CT study of the thorax, showed significant decreasing of the mass

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[1] Computed Tomography.

[2] Multi-Planar Reconstruction.

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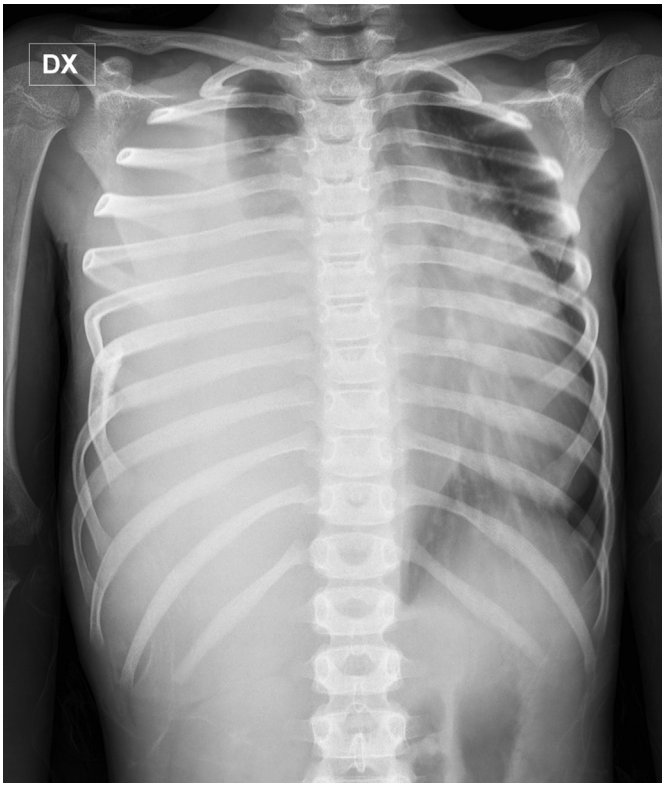

Figure 1. Posterior-anterior chest radiography. It reveals a large opacity which occupies most of right hemithorax with an involvement of the lateral arch of the seventh rib. It appears with thinning of the bone matrix and sclerotic reaction of the cortical bone. Note the displacement of mediastinal structures to the left side and the poor visualization of the right diaphragmatic profile with obliteration of the omolateral costo-phrenic angle.

size and disappearance of the displacement of mediastinal structures (Fig. 3A–D). The patient was a candidate for surgical resection and successively he was subjected to rib resection with right lateral thoracotomy and excision of the mass and the adjacent lung parenchyma.

Discussion

Askin's tumor is an uncommon, malignant, small round cell tumor which belongs to the Ewing's sarcoma family of tumors.[1] More specifically, it is a peripheral primitive neuroectodermal tumor (PNET) located exclusively in the thoracopulmonary region, which was first described in childhood by Askin et al., in 1979.[2] Ewing sarcoma and peripheral Primitive Neuro-Ectodermal Tumors (PNETs[3]) were originally described as distinct clinic-pathologic entities: in 1918, Stout described a tumor of the ulnar nerve composed of small round cells focally arranged as rosettes; this entity was subsequently designated neuroepithelioma, and then PNET.[2] Later, in 1921, Ewing sarcoma was described as an undifferentiated tumor involving the diaphysis of long bones. It was also reported to arise in soft tissue (extra-osseous Ewing sarcoma).[3] Nevertheless, it has become clear, over the last three decades, that these entities comprise the same spectrum of neoplastic diseases known as the Ewing sarcoma family of tumors, which also includes malignant small cell tumor of the chest wall and atypical Ewing sarcoma. Ewing sarcomas of the chest wall were originally reported by Askin in 1979 in 20 white children and adolescents (average age, 14 years).[1] Since then, PNETs within the thoracopulmonary region have been termed Askin tumors. This is a rare disease in the pediatric group, and even rarer in adults. PNETs and Ewing sarcoma are closely related malignant, small, round-cell tumors of soft tissues and bones.[1] Recently, PNETs and Ewing sarcoma have been categorized into a group known as the Ewing family of tumors because of their immunohistochemical, ultrastructural, and molecular similarities. Askin tumors are highly aggressive and metastasize rapidly with poor prognosis. The patients usually present a painful mass of the chest wall, cough, fever, dyspnea, and weight loss. The treatment is an intensive combined therapy including resection, chemo-therapy and radiotherapy. Preoperative chemotherapy is useful to reduce the risk of intraoperative tumor rupture and tumor cell dissemination.[3] The disease is diagnosed by histologic and immunohistochemical analysis. The typical feature is the presence of Homer Wright rosettes.[4] Typical CT findings include a large soft tissue mass arising from pleural effusion and rib involvement. Large lesions often have a heterogeneous appearance, representing hemorrhage and necrosis or cystic degeneration. Calcification within the mass and lymphadenopathy are rare. The mass tends to displace, rather than encase, the adjacent organ. Linear and nodular density in the subpleural fat, which may represent local peri-lymphatic and peri-neural extension, is an unfavorable prognostic sign.[2] After intravenous contrast administration, the tumors demonstrate a heterogeneous appearance. Askin tumor is liable to recur in proximity of the resected tumor, commonly involving the ribs, pleura, chest wall muscles and diaphragm.[5] The metastasis sites include lung, mediastinal and retroperitoneal lymph nodes, extra thoracic skeleton, liver, adrenal glands, and sympathetic nerve chain.[6] Nevertheless, the radiologic characteristics of Askin tumors are not specific,[5,7] but imaging can be useful for evaluating the extent of the tumor, the response to treatment and local recurrence or distant metastasis. It is also valuable for guiding the biopsy route and determining whether the tumor has been completely resected. Askin tumor is in differential diagnosis with neoplasms that can show a mass of chest wall at CT scan, including neuroblastoma, rhabdomyosarcoma, non-Hodgkin lymphoma and Langerhans cell histiocytosis. Neuroblastomas occur before 5 years of age, and characteristically present invasion through neural foramina giving a dumbbell appearance.[8] Rhabdomyosarcoma and Askin tumor at CT imaging, can present similar characteristics, so a biopsy is necessary for diagnosis. Non-Hodgkin lymphoma usually shows nodular thickening of the pleura and rarely isolated chest wall masses. It is important to exclude non-Hodgkin lymphoma as its treatment does not include surgery.[8] Imaging features of Langerhans cell histiocytosis are bony lytic lesions with or without soft tissue masses, with multi-systemic involvement such as brain, lung and abdominal organs. In this case, the CT images show similar characteristics between Askin tumor and others neoplasms that appear as a mass of chest wall. The differential diagnosis need to include also this rare tumor occurring in children because a prompt treatment and an early evaluation of prognosis should be employed.

[3] Primitive Neuro-Ectodermal Tumors.