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ABDOMINOPELVIC INFLAMMATORY MYOFIBROBLASTIC TUMOR DETECTED IN A FIVE MONTH-OLD INFANT: A CASE REPORT

Beş Aylık Bebekte Abdominopelvik İnflamatuar Myofibroblastik Tümör: Olgu Sunumu

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ABSTRACT

The inflammatory myofibroblastic tumor (IMT) is a rare tumor with intermediate potential of malignancy usually encountered in children and young adults and located in lungs, abdominopelvic and retroperitoneal areas. The etiology and pathogenesis of IMT are still uncertain. IMT can mimic various benign or malignant tumors due to absence of specific clinical or radiologic findings. Determination of immunohistochemical ALK positivity can be helpful in differential diagnosis and prediction of prognosis. Herein, we present a case of abdominopelvic IMT detected in a 5 months old girl with preoperative diagnosis of neuroblastoma and discuss its clinical and pathological characteristics.

Key Words: Inflammatory myofibroblastic tumor, abdominopelvic, ALK

ÖZET

İnflamatuar myofibroblastik tümör (İMT), özellikle çocuklar ve genç erişkinlerde görülen, sıklıkla akciğer, abdominopelvik ve retroperitoneal yerleşimli olan intermedier malignite potansiyeline sahip nadir bir tümördür. Etyoloji ve patogenezi halen net anlaşılamamış İMT, spesifik klinik ve radyolojik bulgular içermediğinden pek çok benign ve malign tümörü taklit edebilir. Ayırıcı tanısında ve prognozu öngörmede immunohistokimyasal ALK pozitifliğinin tespiti yol göstericidir. 5 aylık kız bebekte saptanan ve radyolojik olarak nöroblastom olduğu düşünülen abdominopelvik İMT olgumuz klinik ve patolojik özellikleriyle sunulmuştur.

Anahtar Kelimeler: İnflamatuar myofibroblastik tümör, abdominopelvik, ALK

INTRODUCTION

The inflammatory myofibroblastic tumor (IMT) is a rare tumor with an intermediate malignancy potential which was previously called inflammatory pseudotumor, plasma cell granuloma, omental-mesenteric myxoid hamartoma or inflammatory fibrosarcoma (1, 2). It is most commonly located in lungs (3); however, when extrapulmonary located, abdomen, particularly retroperitoneum and mesenterium are the most involved sites (4). Rare visseral locations such as the urinary bladder (5), external auditory canal (6), larynx (7), breast (8), stomach (9), spleen (10) and pancreas (11) have also been reported. IMT, commonly observed within the first decade of life and in young population, may be confused with other benign and malignant tumors clinically and radiologically (11, 12). Therefore, it should be noted for differential diagnosis of intraabdominal and pelvic masses.

Herein, a case presented with abdominal distention and symptoms due to mass effect who was considered as neuroblastoma radiologically but diagnosed as IMT as a result of pathological examination is presented with its clinical and pathological characteristics.

THE CASE REPORT

A 5 month-old female infant was referred to hospital by her parents due to abdominal distention and difficulty in urination and defecation noticed for last 1 month. After detection of a mass of 10x10 cm filling the abdomen by ultrasonographic imaging, she was referred to pediatric surgery department of our hospital for further examination. In the abdominal CT scan,a centrally cystic 10x9.5x6 cm solid mass with peripherally enhanced contrast uptake filling almost all the midline in the pelvic area and extending to bilateral sides of the abdomen and sacral foramina was detected. Due to radiologically detected relation of the lesion with sacral foramina, the patient was operated by a pre-diagnosis of “Neuroblastoma” originating from the sympathetic ganglion. During the surgery, the mass was observed to have a thin capsule adherent to the urinary bladder and to be invaded the anterior wall of the rectum and the sigmoid colon; the mass was excised totally. On gross examination of the mass, it weighted 343 g and was 10.5x9.5x6.5 cm, with an encapsulated outer surface focally irregular appearance and a cream colored, homogeneous cut surface.(Figure 1). On microscopic examination of the sections obtained from the mass, a tumor surrounded by a focally deformed fibrous capsule infiltrating the surrounding striated muscle and fat tissues was observed. The tumor consisted of cells with oval-fusiform nuclei with prominent nucleoli and pale fibrillary cytoplasm which constitutes fascicles and storiform patterns and exhibit focal nuclear atypia (Figure 2). There was a mixed inflammatory cell infiltration including lymphocytes, plasmocytes, and fewer neutrophils and eosinophils within the tumor cells in a haphazard distribution (Figure 3). Stromal edema, hyalinization and necrosis were also observed in some areas. 5 mitosis were detected in 10HPF. Immunohistochemical analysis revealed diffuse positive expressions with ALK, vimentin, SMA, caldesmon, desmin and CD 31 whereas a focal positive expression with Beta catenin and negative expressions with S-100, EMA, Bcl-2, myogenin, CD 99 and CD 117 were detected (Figure4). Ki-67 proliferation index was 20%. In the light of these findings the case was diagnosed as “Inflammatory Myofibroblastic Tumor”. The patient is still under close clinical follow-up after surgery with no additional treatment.

DISCUSSION

IMT is a rare mesenchymal tumor with an unclarified etiology and pathogenesis (5, 8). Possible role of infection, inflammation, surgical procedures, autoimmunity and chromosomal variations have been addressed in development of IMT and localized deviations on chromo-

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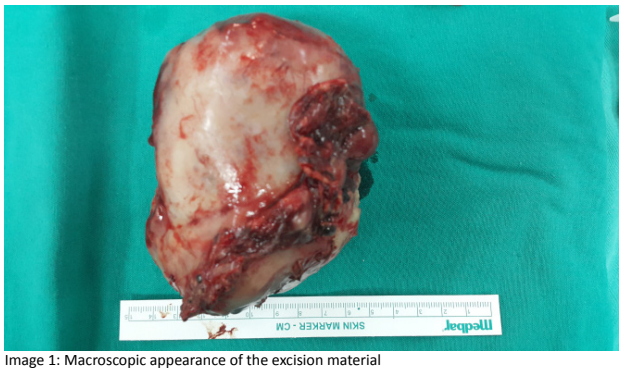

Image 1: Macroscopic appearance of the excision material

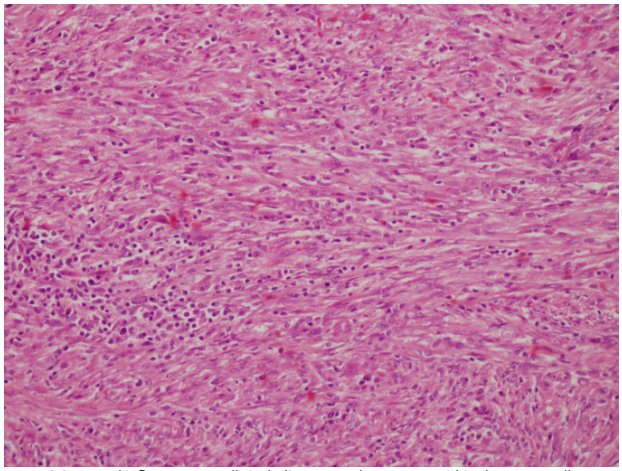
Image 3:Scattered inflammatory cells including many plasmocytes within the tumor cells (HEX40).

somes 2 and 9 has been revealed in some recent studies (5, 10).

IMT which has been first described in 1939 was previously accepted as a benign lesion; however, a local recurrence rate of 37% and a metastasis rate of 11% in the case series reported during last 20 years have revealed a potential aggressive progression of the disease (12). The World Health Organization (WHO) classified IMT as a tumor with an intermediate malignancy and a rare metastasis potential in 2002 (4, 12). Since the tumor may mimic malignancies clinically, radiologically and microscopically, it had been also called inflammatory pseudotumor previously (6).

Average age for IMT is 9 years and it is usually detected in children and young adults; the tumor is slightly more encountered in women than men (10, 12). Our case is a 5 month-old female infant and one of the youngest cases diagnosed as IMT in the literature.

IMT can arise in many different anatomic locations, most commonly in lungs followed by abdominopelvic area and retroperitoneum (5, 6, 12). Tumor diameter varies between 2 and 20 cm and average diameter is 5 to 10 cm (1, 2). Macroscopically IMT appears as lobular or multinodular solid mass with a gray-skin colored or reddish cut surface. It can mimic many benign and malign neoplasms such as leiomyoma, leiomyosarcoma, inflammatory liposarcoma macroscopically (4, 12). Tumors located intraabdominally are usually solid, white colored, irregular masses enlarging inside the mesenterium and are generally adherent to the intestinal wall and sometimes invade it (12). The mass detected in our case was 10.5 cm in diameter with a solid, firm cut surface and focal irregular appearance on the outer surface, located in the pelvic area extending to lateral walls of the abdomen. Furthermore, the mass was adherent to the urinary bladder and invaded the anterior rectal wall and the sigmoid colon. Radiologically detected relation of the mass with sacral foramina caused the suspicion of “Neuroblastoma” originating from the sympathetic ganglion.

Clinical and radiological findings are nonspecific in diagnosis of IMT; therefore, final diagnosis is based on histopathological examination of the excision material (5, 9). Histopathologically, infiltration of the inflammatory cells including plasma cells, lymphocytes and eosinophils are observed along with proliferation of the myofibroblasts (1,

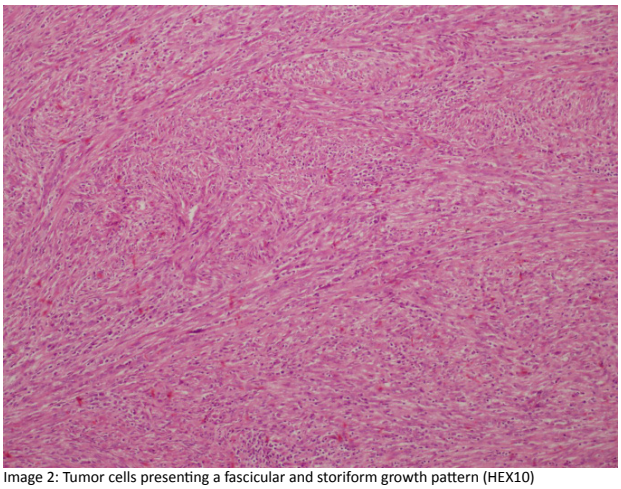
Image 2: Tumor cells presenting a fascicular and storiform growth pattern (HEX10)

12). Presence of the inflammatory cells is an important characteristic allowing differentiation of IMT from fasciitis and fibromatosis (1). The neoplastic myofibroblasts constitute a fascicular or storiform pattern (1, 2). Some cases have a hypocellular pattern and may include hyalinized or collagenous stroma and mimic a scar or a desmoid tumor (1, 2, 12). Necrosis and calcification may be observed rarely (1). Slight focal cellular atypia and mitosis may accompany to the tumor; however, atypical mitosis is not expected (2, 6). A cellular tumor with a fascicular and storiform pattern was observed in our case; focal stromal edema, hyalinization as well as focal necrosis were also detected. Focal atypia and rare mitosis were along with the lesion.

Immunohistochemical analyses play a crucial role indifferential diagnosis of IMT. Positive expression with vimentin, actin and desmin are detected in the tumor cells (2, 6, 12, 13). Role of a mutation on anaplastic receptor tyrozine kinase gene located on the short arm of chromosome 2 has been shown in the pathogenesis of the disease and by immunohistochemical staining, cytoplasmic positivity can be detected with anaplastic lymphoma kinase (ALK) in approximately 50% of the cases (7, 8, 12, 13). ALK positivity can be used to differentiate IMT from ALK-negative tumors such as leiomyoma, leiomyosarcoma, fibromatosis, nodular fasciitis, calcified fibrous tumor, myofibromatosis and infantile fibrosarcoma (12, 13).

Moreover, some studies revealed that ALK-negative IMTs present a more aggressive clinical course due to increased risk of metastases and local recurrence (1, 6, 9, 12, 13). Thus, detection of ALK with immunohistochemical method is suggested to be beneficial in prediction of prognosis.

Clinical behavior of the tumor also differs depending on the location. Primary IMTs with abdominopelvic location tend to cause increased local recurrence with a rate of 25% compared to primary pulmonary IMTs with a local recurrence rate of 2% (2, 12). Among the most common sites for metastasis are lungs, mesenterium and omentum (12).

Surgical excision is curative for treatment of IMT. Therefore, differentiation from sarcomas and making accurate diagnosis are quite important to prevent unnecessary aggressive therapies (9). Moreover, detection of ALK expression is critical to direct the patients for an adjuvant therapy (12). More aggressive therapy regimens may be required for ALK-negative patients (13).

Adjuvant therapy has not been considered in our case by virtue of ALK positivity; however, the patient is under close follow-up clinically due to abdominopelvic location, large tumor size and invasion of the tumor to adjacent organs.

In conlusion, IMT is an entity which should be considered in differential diagnosis of intraabdominal and pelvic located masses, especially during childhood. The final diagnosis is possible through an histopathological examination because the tumor can mimic many different benign and malign tumors clinically and radiologically. Immunohistochemical stains are used for differential diagnosis, as well as positive expression of ALK, is very important to plan the treatment and predict the prognosis.

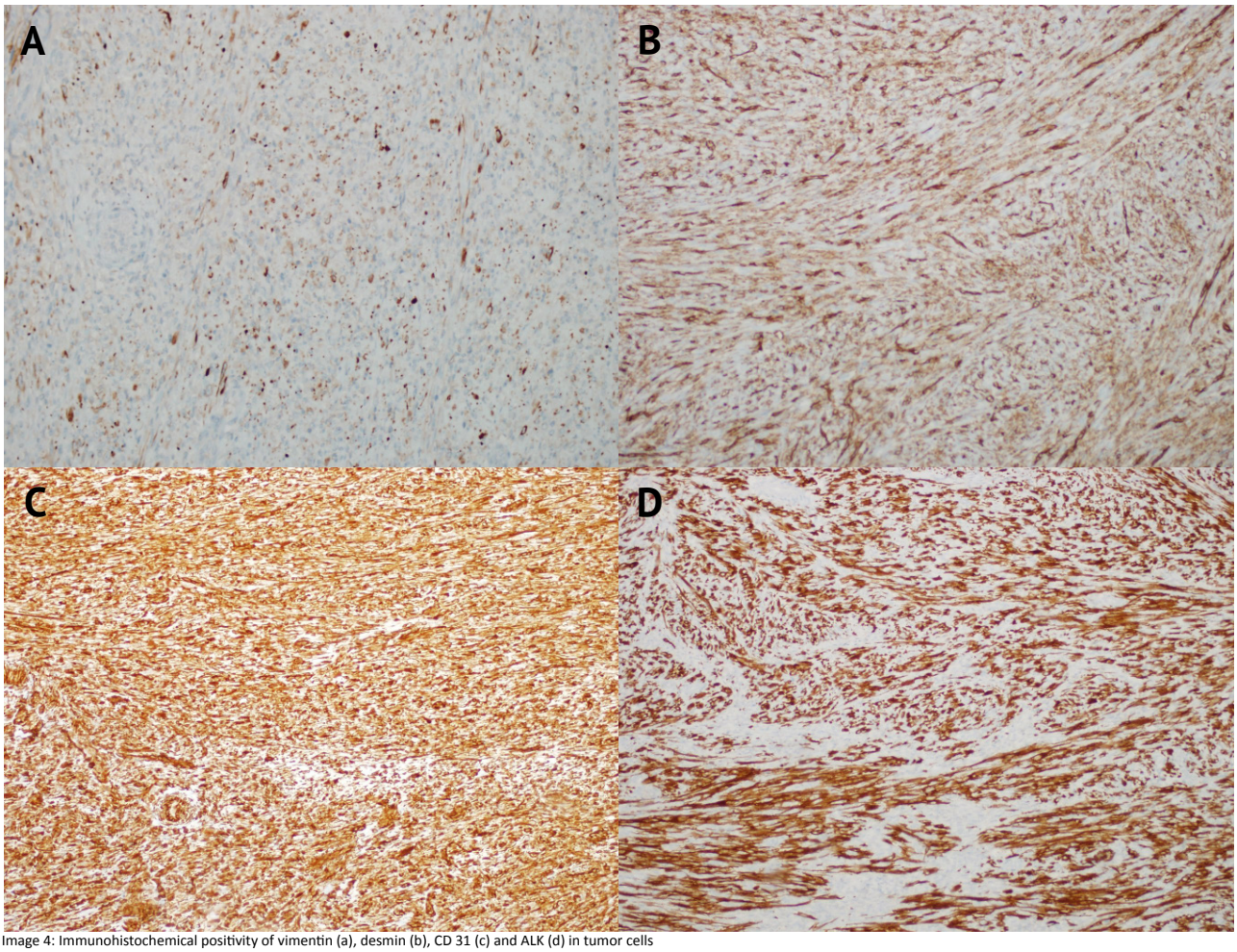

Image 4: Immunohistochemical positivity of vimentin (a), desmin (b), CD 31 (c) and ALK (d) in tumor cells

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Conflict of Interest

The authors declare no conflict of interest.