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Case Report

Fatal Outcome of Recurrent Infantile Pelvic Desmoid Tumor Treated with Tamoxifene

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Abstract

Desmoid tumors are rare benign neoplasms with an aggressive local growth. In children, intra-abdominal localization is less frequent and few reports exist in the literature about the management of DTs in those special patients. In our report, we describe a case of a 13–year old patient with a bifocal intra–abdominal DT, treated unsuccessfully with tamoxifene, and we discuss briefly the existing literature data.

Keywords: infantile desmoid tumor, Recurrence, Tamoxifene, intra–abdominal site.

Introduction

Desmoids tumors are rare benign tumors of fibroplastic origin with a strong tendency to invade locally and to recur. They constitute 3% of all soft tissue tumors and 0.03% of all neoplasms (1). Although desmoid tumors are more common in the second to fifth decades of life, they can appear in younger children and older adults. In pediatric patients, desmoid tumors occur rarely in the pelvis. In this paper, we report arare case of pelvic desmoid tumor in a 13–year–old girl treated with tamoxifene who experienced fatal outcome.

Case Report

A 13–year–old girl presented to our hospital for a painless distension of the abdomen for five months. She had a medical history of surgery for ovarian fibrothecoma one year ago. Physical examination revealed cachexia with a painless distended abdomen. Enhanced Computed tomography scan of abdomen and pelvis showed a large neoplastic in homogeneous abdominopelvic mass without involvement of adjacent structures. No evidence of tumor origin was detected. AL2 – S2 paravertebral mass with the same characteristics was also seen. Magnetic resonance imaging (MRI) demonstrated an irregular lobulated solid mass in peritoneal cavity measuring 14cm x 20cm x 15.3cm, extending to anterolateral abdominal wall, anterior surfaces of L4, L5 vertebrae and displacing bowel loops and bladder, without signs of locoregional involvement (Figure 1 a,b,c). A secondary right paravertebral mass4.8cm x 5.6cm x 11cm in size, extending from L2 to S2 without involvement of bone structures was individualized (Figure 1d). Both lesions showed low signal intensity T1, iso – intensity T1 and heterogenous enhancement after Gadolinium injection. No evidence of hepatic or bone lesion was seen. The MRI imaging features were suggestive of soft tissue sarcoma with paravertebral secondary localization.

The previous specimen of resected ovarian fibrothecoma was reexamined, and the histopathological analysis was found to be compatible with a desmoid tumor invading the ovary. Core needle biopsy of both lesions was then performed. Histopathological examination confirmed the diagnosis of bifocal desmoid tumor (Figure 2). Complete surgical resection with wide margin could not be performed because of the depth of paravertebral mass. Thus, the patient received tamoxifene at a dose of 20mg twice daily. The clinical course of patient was unfavorable and she succumbed to her disease three months after the initiation of medical treatment.

Discussion

Desmoids tumors (DTs) or aggressive fibromatosis are rare benign neoplasms from musculoaponeurotic origin, with an aggressive local infiltrating growth, without any tendency of metastasis. DTs affect all age groups. The most frequent anatomic sites of DTs in children are extremities...
and head and neck; whereas abdominal locations are rare accounting for 5 to 7% of disease in children\(^2\,^3\). Some etiologic factors have been reported in the literature such as trauma to the region including surgical trauma\(^3\), high estrogenic states \(^4\), and hereditary cancer syndrome\(^5\). In most cases, DTs occur sporadically, a \(\beta\)-catenin activating mutation is more common in this setting\(^6\). Most of the intra-abdominal DTs are asymptomatic until complications due to local invasion or compression of surrounding structures occur. Patients with intra-abdominal DTs present typically with a painless enlarging mass. Severe complications like bowel obstruction, hydronephrosis and cachexia have been described\(^8\,^9\). Like in our case, DT might be synchronous or multifocal in 6% of patients\(^2\,^7\). Magnetic resonance imaging (MRI) is the most appropriate imaging modality to diagnose and to evaluate tumor origin and regional extent of disease. Characteristics of MRI findings show low signal intensity relative to muscle on T1–weighted images, and a variable signal intensity on T2–weighted images \(^12\). Histologically, DTs appear with well–differentiated cells and bands of fibrous tissue; margins can be difficult to identify \(^22\). Gross total resection of the tumor with negative margins is the mainstay of treatment of DT\(^22\). Adjacent structures involved by tumor should be also removed. Incomplete tumor resection or positive surgical margins may impact negatively event–free survival \(^13\,^15\). However, radical surgery would be mutilating or associated with considerable function loss\(^16\). Alternative therapeutic approaches, such as radiotherapy, chemotherapy, endocrine therapy and nonsteroidal anti–inflammatory agents could be proposed in those cases as well as in the setting of residual, recurrent, progressive or unresectable tumor, with variable success \(^17\,^18\). Because of our patient’s young age and the potential adverse effects, we opted for hormone therapy. Endocrine therapy including
tamoxifene have shown anti-tumoral activity in DTs, with response rates of over 50% have been reported in adults (19). However the efficacy of hormone based therapies has not been well proven in pediatric patients, and their impact on the growth and development of young patients is unknown (20). Combination of hormone therapy with chemotherapy might be beneficial but long term clinical response is minimal (21). In our case, tamoxifene failed to attempt any clinical response; maybe a combination of two therapeutic options would be more efficient as the course of disease was not indolent (short period between initial resection and relapse, bifocal extensive disease).

Conclusion

DTs exhibit worse prognosis because of their aggressive growth and local invasion. Several therapeutic approaches are available with variable results. There is no standard approach in the management of recurrent tumors especially in pediatric patients. Our case highlights the importance of clinical trials to define the best therapeutic approach of these patients especially for unresectable tumors.

References


Figure 2: Proliferation of well differentiated spindle cells arranged in moderately rich collagen–dense arrays (a=HEX100). There was no mitosis or necrosis (b=HEX400).


