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CASE REPORT OF A TWO YEAR OLD BOY WITH DESMOID TUMOR OF RECTUS ABDOMINIS

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Abstract:

The other name of desmoid tumor is aggressive fibromatosis, locally aggressive injury to soft tissues caused by connective soft tissue. Reports are rare in children under 10 years of age. We present a desmoid tumor case in a 2-year-old boy at the middle and lower third of the left rectus abdominis. Reconstruction of the abdominal wall with abdominal fascia were performed and partial resection of the muscle segment was done simultaneously. The patient recovered without incident. There was no recurrence, functional or aesthetic complication at the end of one year follow-up.

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INTRODUCTION:

Desmoid tumors, also called as aggressive fibromatosis or desmoid fibromatosis, are benign neoplasms of mesenchyme characterized by aggressive local infiltration of surrounding tissues with no growth and metastatic potential, but with the chance of recurrent growth and greater recurrence [1-3]. Although described by MacFarlane in 1832, the term desmoid was first given in 1838 by Muller in Berlin, who used the term "desmos" for the Greek word - "tendon-like". Desmoid tumor is a rare, deep muscle-aponeurotic moderate malignancy tumor with an incidence of 0.2 to 0.4 per 100,000 people per year [4-6]. These are slow-growing tumors and it takes years to get an extraordinary mass [7-9]. Infants are rarely reported. Here, we report an anterior abdominal wall desmoid tumour in the baby.

CASE REPORT:

A two year old boy came to the hospital with two months history of progressive increase in mass at paraumbilical region. There was no history of trauma and the baby was otherwise normal. The patient guardians definite to attain medical management for discomfort in the abdominal. Neonatal history, birth and pregnancy history was unremarkable and accomplished the National Vaccination Program according to age. General physical examination was normal. Physical examination revealed a painless supra and umbilical mass measuring 4 x 2 cm in diameter in the left abdomen with well-defined edges, measuring the spherical shape. The mass was soft, hard in nature and not devoted to the skin, but in underlying structures, it was attached. Rectal examination was normal essentially. Magnetic resonance imaging (MRI) and CT scan of the abdominal region shows a heterogeneous, ovoid mass of 43 x 12 x 20 mm limited to the sheath of the rectus abdominis and located in the middle and lower three of the abdominal rectus muscle (Figure 1A). The operation was performed with middle and lower rectus muscle resection and completely removed. The defect was covered with the surrounding fascia. A histopathological analysis established the desmoid tumor diagnosis with free of tumour surgical margins (Figures 1B, 1C, ID). The patient recovered without incident. No recurrence was observed at the end of one year follow-up.

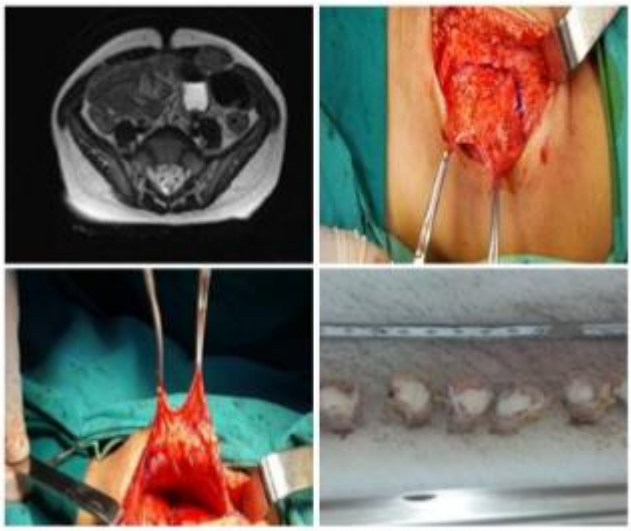

Figure 1: (A): Magnetic resonance showing ovoid lesion and hypodense mass in left rectus abdominis sheath; (B,C): Perioperative picture; (D): Cut section of the mass.

DISCUSSION:

Desmoid tumors represent 3% of all soft tissue tumors. They are classically slow-growing mesenchymal tissue; however, they may penetrate in to the deeper tissue planes, therefore, the higher the chances of repetition. They usually occur in the 2nd or 3rd span of life, but various succession have recognized desmoid tumors in elderly or middle-aged persons [10-11].

In 1st epoch of life; desmoid tumor occurrence is very uncommon, this case was 2 years old and for about 2 months had detected anterior abdominal mass but asymptomatic and had only moderately amplified in size due to the slow growth nature of the tumor. Surgery revealed a well-constrained tumor involved in the rectus abdominis muscle at its inner layer and was completely excised with a tumor margin of approximately 1 cm. In children, a negative resection

margin significantly reduces tumor recurrence, but in adults, recurrence may occur at tumor-free margins [12-13]. The diagnosis is based on clinical, radiological and histopathological criteria. Its main feature is the penetration of muscles and deep structures. This characteristic distinguishes the desmoid tumor from sarcomas. It is most commonly seen in persons with (FAP) familial adenomatous polyposis, which occurs as hereditary syndrome (Gardner syndrome) [14]. From myofibroblasts; desmoids are arise, without a real capsule, and mostly penetrate the nearby muscles. The activity and length of telomerase was usual, the nuclei were well-regulated and small, and mitosis is rare, all supporting histologically benign nature. Surgery is the basis for the treatment of desmoid tumors in the abdominal wall. Large local resection, which has been advocated for almost 100 years ago, rests the 1st line of treatment for desmoid tumors patients [15]. With 1 cm free surgical margins; excision should be done and the with local muscle flaps defect should be revamped, distant muscle flaps, or if essential synthetic mesh may be applied. If operated properly, relapse rates are low and procedural morbidity rates are negligible. Historically, negative border resection has been the gold standard for the treatment of abdominal wall and intraabdominal desmoid tumors.

CONCLUSION:

In conclusion, it has been discovered that initial surgery with free tumour margins is the prime successful management for children with desmoid tumors. Long-term follow-up is required to select any recurrence.

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