

Intra-abdominal Desmoid Tumor Presenting as Bowel Obstruction in a Child with Multiple Congenital Abnormalities

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Citation

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Abstract

Intra-abdominal desmoid tumors are rare soft tissue tumors. We describe a young male child with a sporadic intra-abdominal desmoid tumor presenting with features of acute intestinal obstruction. Such a presentation is extremely rare and only a few such cases have ever been reported.

CASE REPORT

A male child aged 14 years arrived in the emergency room of SMHS Hospital with a history of abdominal pain, distension, vomiting and constipation of 3 days' duration. There was no history of fever, trauma, previous abdominal surgery or passage of worms with stools. There had been no similar history in the past and the family history was not significant. Abdominal examination revealed features of intestinal obstruction and a vague fullness was palpable in the left upper quadrant of the abdomen. On general physical examination, the patient had left sided facial asymmetry and polydactyly. Investigations revealed leucocytosis, multiple air-fluid levels on standing abdominal x-ray and distended gut loops as well as an absent left kidney on abdominal ultrasonography.

A diagnosis of acute intestinal obstruction was made and an emergency surgical exploration was performed after conservative measures had failed. On laparotomy, the small gut and the right half of the transverse colon were found to be distended. There was a mass wrapped by omentum pressing upon the splenic flexure. On dissection, it was found to arise from the parietal wall and was completely enucleated. Intrigued by the patient's multiple congenital abnormalities and the conspicuous absence of the left kidney on ultrasound, we conducted further investigations postoperatively including abdominal CECT, which confirmed the absence of the left kidney, and colonoscopy, which was normal. Also, the radiographs of the skull confirmed hypoplasia of the left maxilla. The

histopathological study of the surgical specimen revealed desmoid pathology. The patient's entire clinical picture did not fit into any of the recognized clinical syndromes. After confirmation of the diagnosis; since a complete curative resection was not undertaken, the patient was put on chemo-radiotherapy and is on follow-up for detection of any recurrence.

Figure 1



Figure 2



Figure 3



Figure 4

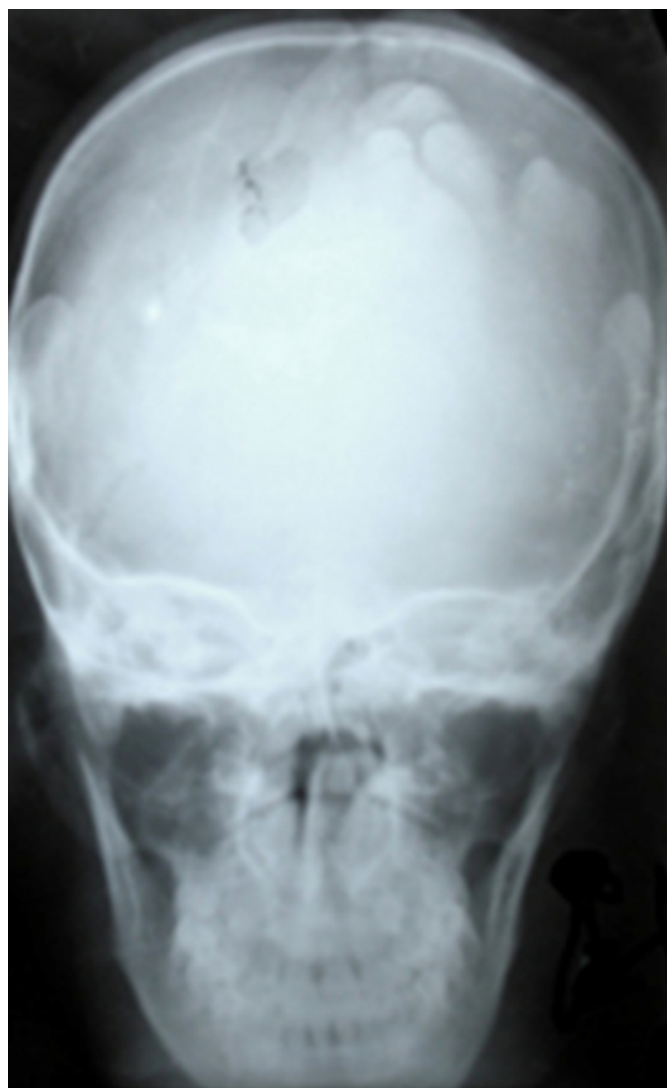
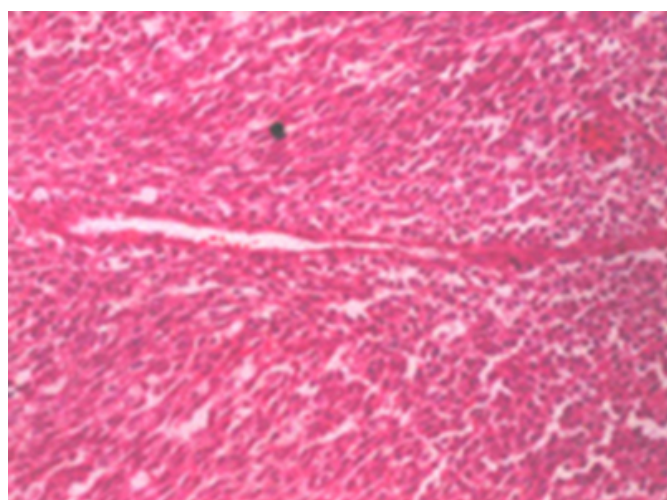


Figure 5



DISCUSSION

Desmoid tumors are rare, benign slowly growing tumors and account for 0.035% of all neoplasms and less than 3% of all soft tissue tumors. Incidence rate is 2-4 per million per year.¹ Despite their benign appearance they are locally aggressive & invasive but do not metastasize.² The etiology is unknown. A connective tissue growth disorder has been implicated and the tumor is commonly associated with Gardner's syndrome, abdominal surgery and trauma,² pregnancy, estrogen therapy and Crohn's disease. However, in children these associations are difficult to establish. Desmoid tumors arise from musculo-aponeurotic tissue and two forms are distinguished – intra-abdominal and extra-abdominal. The former is rare and arises from the abdominal wall, less commonly from the mesentery and retroperitoneum. The common extra-abdominal locations include the limb girdles and buttocks.³ In 10%, multicentric disease has been reported.⁴ Since desmoid tumors are slowly growing, most patients are asymptomatic and so remain undiagnosed. By the time the diagnosis is made, the tumor is usually too large, producing mass effects, and has already invaded neighboring structures causing significant morbidity and complications. Tumors often recur after surgery (recurrence rate 75-86%). Distant hematogenous and lymph node metastasis are not seen. The imaging appearance of these tumors is variable and depends on fibroblastic proliferation, collagen content and vascularity. On ultrasound, they appear as masses of variable echogenicity with smooth well-defined margins.⁵ On CT, most appear as well-circumscribed homogenous masses that may be isodense or hyperdense relative to muscle. On MRI, desmoid tumors appear as masses of low and variable signal density relative to muscle, on T1- and T2-weighted images, respectively.⁵ Diagnosis is established by needle or surgical biopsy. Microscopically, they are poorly circumscribed and infiltrate the surrounding tissue. They are composed of spindle- or stellate-shaped fibroblastic cells embedded in a collagenous stroma.⁶ The cells lack atypia and the nuclei are small and pale staining. Surgical treatment is indicated in symptomatic patients or when there is a risk to adjacent structures, in which case wide surgical excision is the procedure of choice, given the locally invasive nature of the lesion and its high recurrence rate. Other modalities of treatment include low dose radiation therapy,² and chemotherapy, including non-cytotoxic agents like NSAIDs (Sulindac), hormonal drugs (Tamoxifen),⁷ and cytotoxic agents which are prescribed when a non-cytotoxic regimen

fails (e.g. combination of methotrexate and vinblastine). Our patient demonstrated a rare presentation of sporadic intra-abdominal desmoid tumor coupled with multiple congenital anomalies, which to the best of our knowledge, did not match any of the known clinical syndromes. The bottom line is that – even though a complete curative resection is the treatment of choice – since our patient was operated under emergent conditions with a clinical diagnosis of acute intestinal obstruction, a complete curative resection could not be undertaken at the time of laparotomy in view of the non-availability of the histological diagnosis. Therefore, in a situation like ours where pre-operative tissue diagnosis is not readily available, second look surgery followed by chemo-radiotherapy or second look surgery with neo-adjuvant therapy appears to be the most viable option.

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