Desmoid-Type Fibromatosis in a Young Male

Fibromatose do Tipo Desmóide num Jovem

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A 17-year-old boy presented to our emergency department with an 8-month history of pain and swelling in the left buttock. Physical examination revealed a firm, fixed, large mass, with extension to the thigh with no inflammatory signs. Magnetic resonance imaging (MRI) showed a mass measuring 17.1 x 14.1 x 10.3 cm (Fig. 1) located deep within the gluteus maximus muscle and adjacent to the sciatic nerve. The biopsy confirmed the diagnosis of desmoid-type fibromatosis (DTF). He started chemotherapy with metronomic vinorelbine (90 mg per week) with stable disease.

DTF is poorly understood and rare in childhood. It is a low-grade malignancy with a high potential for local recurrence. However, it does not metastasize. DTF is commonly seen in the extremities, abdominal wall and mesentery. The definite diagnosis is made histologically. Computed tomography or MRI is needed to assess resectability. Chemotherapy is used in progressive/symptomatic cases where surgery would cause unacceptable morbidity.

AUTHORS CONTRIBUTION
ATR: Draft of the manuscript.
JM, MHA: Critical review of the paper.

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REFERENCES