Currarino syndrome

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A 58-week-old girl presented with chronic constipation. MRI demonstrated sacral dysgenesis, a low-lying conus and a small syrinx (Figs. 1 and 2). Protruding through a ventral sacral defect was a heterogenous fatty mass with intralesional nodules (Fig. 1). Interval nodular enlargement and elevated alpha-fetoprotein (AFP) led to resection of the mass, due to concern for a malignant teratoma. The lesion was proved to be pathologically benign dermoid. The patient had pelvic floor abnormalities and an HLXB9 gene deletion on the 7q36 locus, consistent with Currarino syndrome.

The Currarino triad, as first described in 1981, consists of 1) an anorectal malformation, 2) an anterior sacral defect and 3) a presacral mass, which may be a teratoma, meningocele, neurenteric cyst or combined lesion [1]. Currarino syndrome includes clinical findings such as a bowel obstruction, along with gynecological and renal malformations. Mass resection is advised due to organ compression and the risk of malignant degeneration [2].

References

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Fig. 1 Sagittal T1-W lumbar spine

Fig. 2 Sagittal T2-W lumbar spine