Castleman's Disease presenting as incarcerated femoral hernia in a 4 year old girl

Dear Editor,

We report a case of a 4-year-old girl who presented with a hard, indolent mass which was situated in the left inguinofemoral region. The mass was well demarcated on palpation and although it was painless an incarcerated femoral hernia was suspected initially. Abdominal Ultrasound Scan (USS) showed a circumscribed soft tissue mass, located in the left femoral region and MRI scan a round clearly defined mass of soft tissue with marked enhancement by the contrast medium, situated in the lower left inguinal canal, with dimensions 2.5 x 2.3 cm. Hematology, biochemistry and liver function tests, as well as, C-Reactive Protein, Carcinoembryonic Antigen and A-Fetoprotein levels were found normal. Blunt dissection of the mass was carried out easily through a low inguinal incision. Overall pathology and immunohistochemistry findings were suggestive of hyaline vascular unicentric Castleman's disease. The patient had an uneventful postoperative course. Up to date, 1 year after the operation, follow up studies are normal.

Castleman's Disease (CD) is a rare lymphoproliferative disorder. It has been encountered in every anatomical location of lymph nodes, since it was first described in 1956[1-2]. It appears in two main pathological subtypes: the hyaline vascular (HV) type which is commoner and benign; the plasma cell (PC) type which is rarer with a frequently malignant behavior[3]. Clinically it appears as localized, multifocal-systemic and mixed type[2-3]. CD is infrequent in children and it is most often unicentric with equally divided HV and PC subtypes[3]. The etiology of the disease is uncertain and it is associated with infections including HIV and HHV-8 viruses, and several cancers[4]. CD can mimic a wide spectrum of pathology like autoimmune diseases and malignancies[1-4]. In our patient, a single painless engorged lymphnode was mimicking a femoral hernia which is a unique mode of presentation of CD in childhood. Differential diagnosis can undergo subtraction with USS and enhanced CT or MRI. In children because of the benign nature of the unifocal CD, surgical excision of the affected lymph nodes is the treatment of choice. Chemotherapy, immunotherapy and plasmapheresis are reserved for the rarely unresectable multifocal PC type in children[3]. In conclusion, a higher index of suspicion of Castleman's disease is required for early diagnosis, thus avoiding an unnecessarily prolonged diagnostic workup with a consequent delay of treatment.

References

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Conflict of interest

None declared.

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