PARANEOPLASTIC JUVENILE IDIOPATHIC ARTHRITIS AS A MANIFESTATION OF A BENIGN TERATOMA IN A FIVE-YEAR-OLD GIRL

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Keywords: teratoma; benign; arthritis; pediatric

Abbreviations:

Juvenile idiopathic arthritis	JIA
Non-steroidal anti-inflammatory drug	NSAID
Magnetic resonance imaging	MRI
Computed tomography	CT

Meeting Abstract: Ovarian Teratoma masquerading as Idiopathic Juvenile Arthritis in a 5 year old girl. Presented at the American Society of Pediatric Hematology/Oncology (ASPHO) Annual Meeting. Fort Worth, TX. May 10 - 13, 2023. Poster# 225. 2023 ASPHO Conference Paper and Poster Index. Pediatric Blood Cancer, 70: e30390. S101 https://doi.org/10.1002/pbc.30390

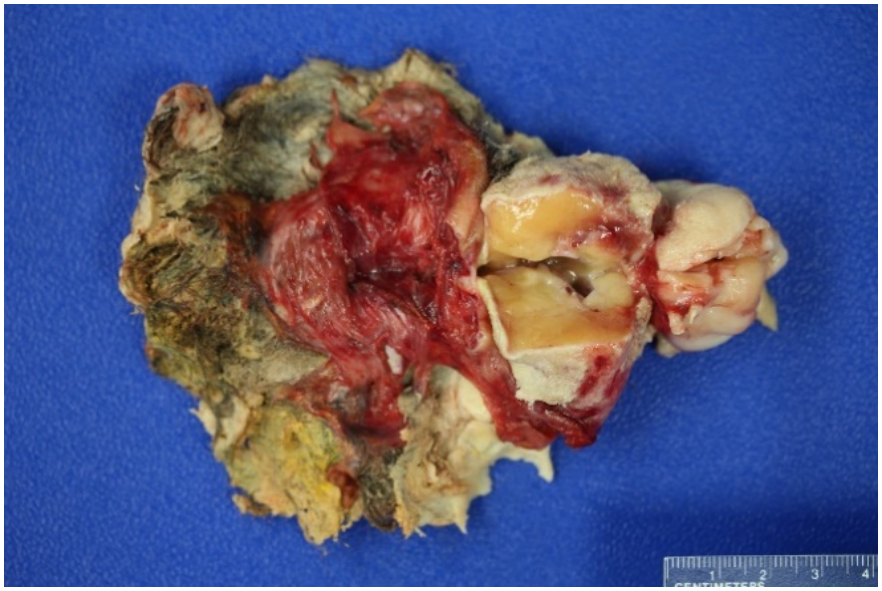
Juvenile idiopathic arthritis (JIA) is an auto-inflammatory disorder that is the most common type of arthritis in children under the age of 16. Thirty percent of patients with JIA have seronegative polyarticular disease. In adults, there are few reports of seronegative polyarthritis associated with ovarian teratomas and sacrococcygeal teratomas; however, to date, there are none reported in the pediatric population.[2,3,4]

A 4-year-old Caucasian female presented with 6 month-history of upper and lower extremity joint pain and swelling associated with myalgias. Family history of psoriatic arthritis, rheumatoid arthritis, and lupus was reported. On examination, she had hypermobility in multiple joints associated with swelling particularly in bilateral knees, right wrist, and left elbow. Complete blood counts, comprehensive chemistry, lactate dehydrogenase, and uric acid were normal. Initial erythrocyte sedimentation rate was elevated at 15 mm/h. Antinuclear antibody and human leukocyte antigen B27 markers were negative. The patient was diagnosed with oligoarticular JIA and started on scheduled non-steroidal anti-inflammatory drug (NSAID) therapy. Due to persistent bilateral knee pain for 8 months despite NSAIDs, magnetic resonance imaging (MRI) of bilateral knees was ordered to look for radiological inflammation or erosive joint disease prior to starting disease modifying anti-rheumatic drugs. As part of routine MRI screening, the mother reported that the child may have swallowed a metallic object, prompting a chest X-ray, which showed a lobular mass with unusual calcifications in the epigastric area. Further investigation prompted computed tomography (CT) and MRI of the abdomen, which showed an epigastric well-circumcised mass (8.8 x 8.3 x 5.5 cm) with soft tissue, fat, and calcified components including numerous tooth-like structures most consistent with teratoma. The MRI of bilateral knees was negative for active inflammation. Tumor markers alpha-fetoprotein and total beta-human chorionic gonadotropin were negative. Tumor resection and histopathology confirmed benign mature retroperitoneal teratoma without immature or malignant components. After tumor removal, the patient's joint pain resolved.

The spontaneous resolution of an inflammatory seronegative arthritis after the excision of the tumoral mass suggests a cause-and-effect relationship between paraneoplastic manifestations and a benign teratoma. It is unclear if the early manifestations of JIA resulted from a genetic predisposition to autoimmune disorders as suggested by the family history.

Teratomas are often found incidentally and most are asymptomatic. There is no known association of JIA with benign teratoma; however, there are abdominal tumors associated with other autoimmune conditions such as juvenile dermatomyositis.[1] This case highlights the importance of thorough clinical evaluation in patients with seronegative arthritis and consideration for teratoma.

Figure 1 Figure 2



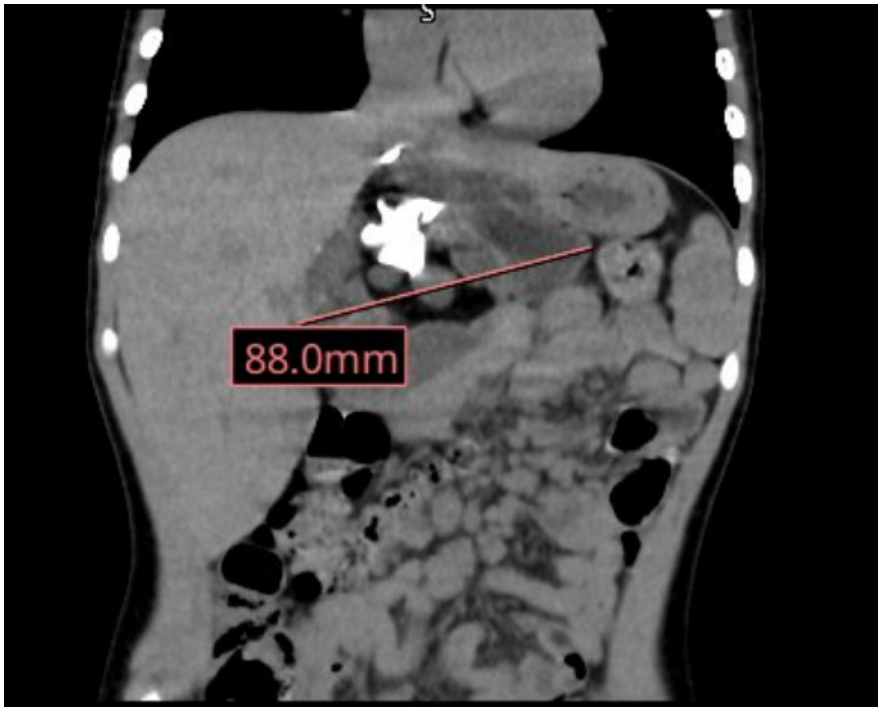


Figure 1: Gross pathology of excised retroperitoneal teratoma, consisting of soft tissue, bone, teeth, fat, hair, and other calcified components

Figure 2: MRI abdomen showed teratomatous lesion extending for hepatic vein confluence superiorly descending into the upper abdomen in the porta hepatis region

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