Abdominal Inflammatory Pseudotumors In The Pediatric Population – A Single Institution's Experience

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Introduction

Background and aim

Abdominal inflammatory pseudotumors are

- Rare
- Variable
- Unpredictable
- Related to ALK gene abnormalities in IMTs

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Introduction

Background and aim

- Aim : To describe our local experience and outcomes with pediatric abdominal inflammatory pseudotumors

Material and Methods

- Retrospective
- Clinical charts of children treated for abdominal inflammatory pseudotumours between Jan 1 2003 – Sept 30 2010 reviewed

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Study Factors

Demographics	Treatment Modalities
Age	
Sex	
Clinical Presentation	**Tumor Pathology**
	Location
Radiological Findings	ALK gene mutations
	Ganglion-like cells
	Outcome

Results

Demographics

No	Age at diagnosis (years)	Sex
1	6	F
2	0.4	M
3	14	F
4	0.5	M
5	11	F

Median age 6 years

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Results

Clinical Presentation

No	Presentation
1	**Intussusception**
2	Incidental abdominal mass, upper chest wall mass
3	**Intestinal obstruction**
4	Incidental abdominal mass, fever
5	Incidental abdominal mass

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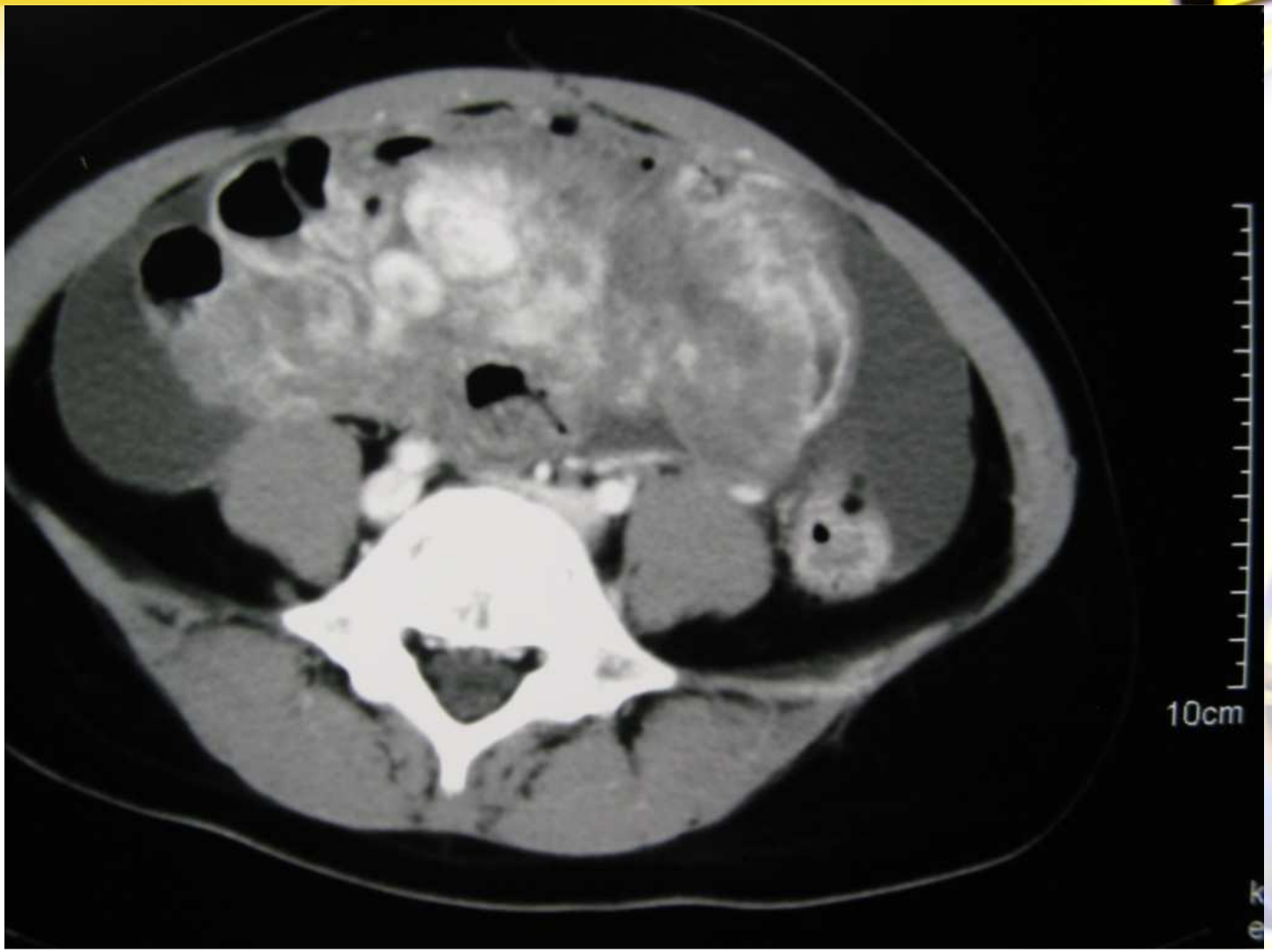
Results

Radiological Findings

No	Location of tumor	Radiological findings
1	Jejenum	US: Target lesion in probably ileocolic region likely intussusception. Hyperechoic nodule likely pathological lead point
2	Mesentery	CT: Large heterogenous lesion 6.5 x 6.5 x 6.5 cm from inferior pole spleen with calcifications
3	Peritoneum, mesentery	CT: Ill defined mass 15cm within lower abdominal mesentery causing intestinal obstruction. Omental nodularity. Ascites
4	Mesentery of sigmoid colon and greater omentum	CT: Large mixed density intraperitoneal mass in right side abdomen 8.2 x 8.8 x 11.4 cm
5	Retroperitoneal and small bowel mesentery	CT: Encapsulated 7 cm retroperitoneal mass with calcifications

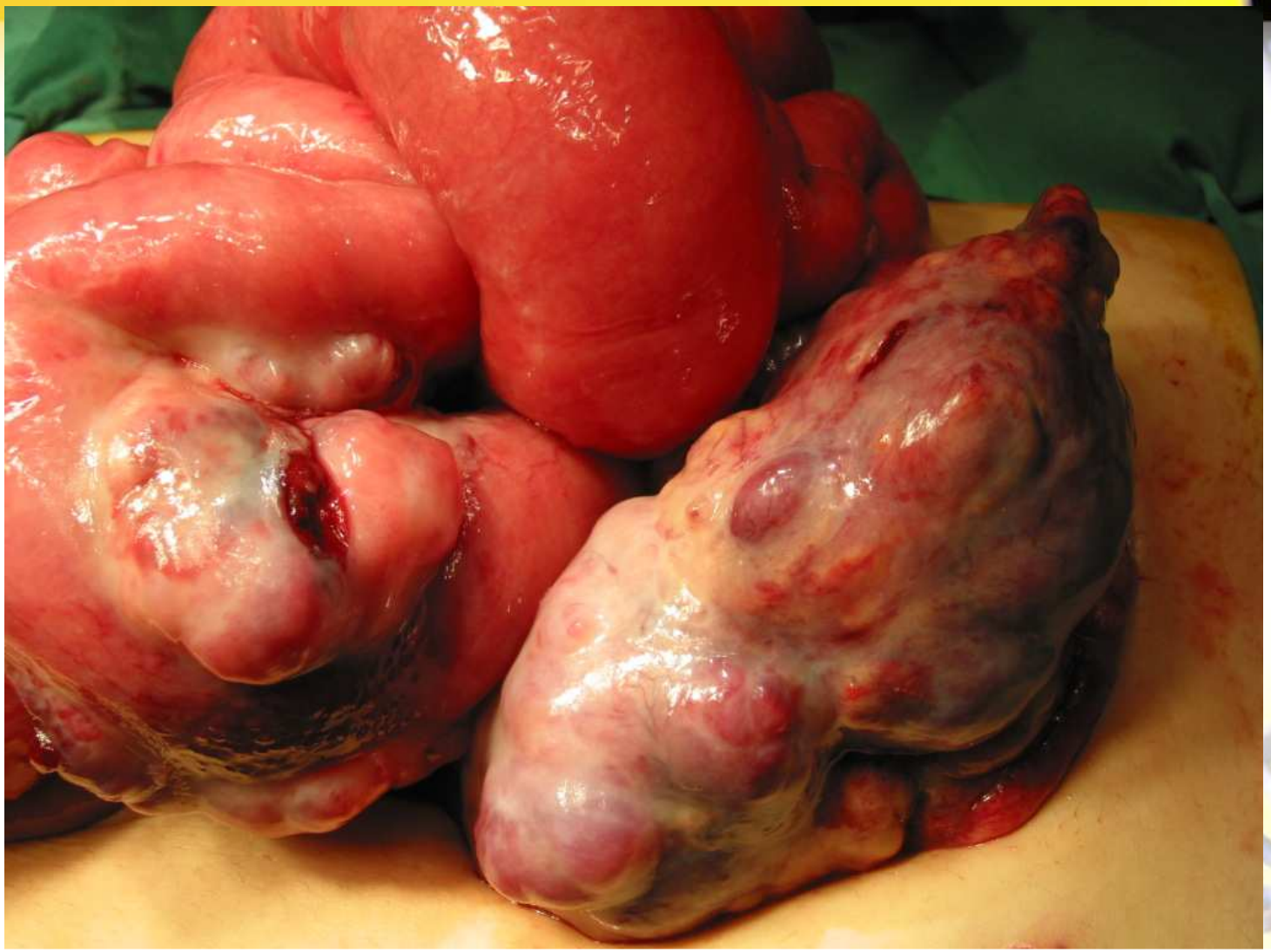
Results

Radiological Findings



Case 3

Results



Case 3

Results

Primary Treatment

No	Surgical Treatment	Histology	ALK gene mutation	Ganglion-like cells
1	**Gross Total resection**	IMT	Not done	Not Present
2	**Debulking**	IMT	Equivocal	Not present
3	**Debulking**	IMT	**Present**	**Present**
4	**Gross Total resection**	IMT	**Present**	**Present**
5	**Gross Total resection**	Calcifying fibrous tumor	Not present	Not present

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Results

Adjuvant Treatment and Outcome

No	Surgical Treatment	Adjuvant	Follow-up period (months)	Outcome
1	**Gross Total resection**	-	14	Disease free
2	**Debulking**	Diclofenac	84	Multi-systemic disease
3	**Debulking**	Diclofenac, chemotherapy	7	Progression of disease➡ Deceased
4	**Gross Total resection**	Diclofenac	10	Recurrence in 4 months➡ resection ➡ Disease free
5	**Gross Total resection**	-	10	Disease free

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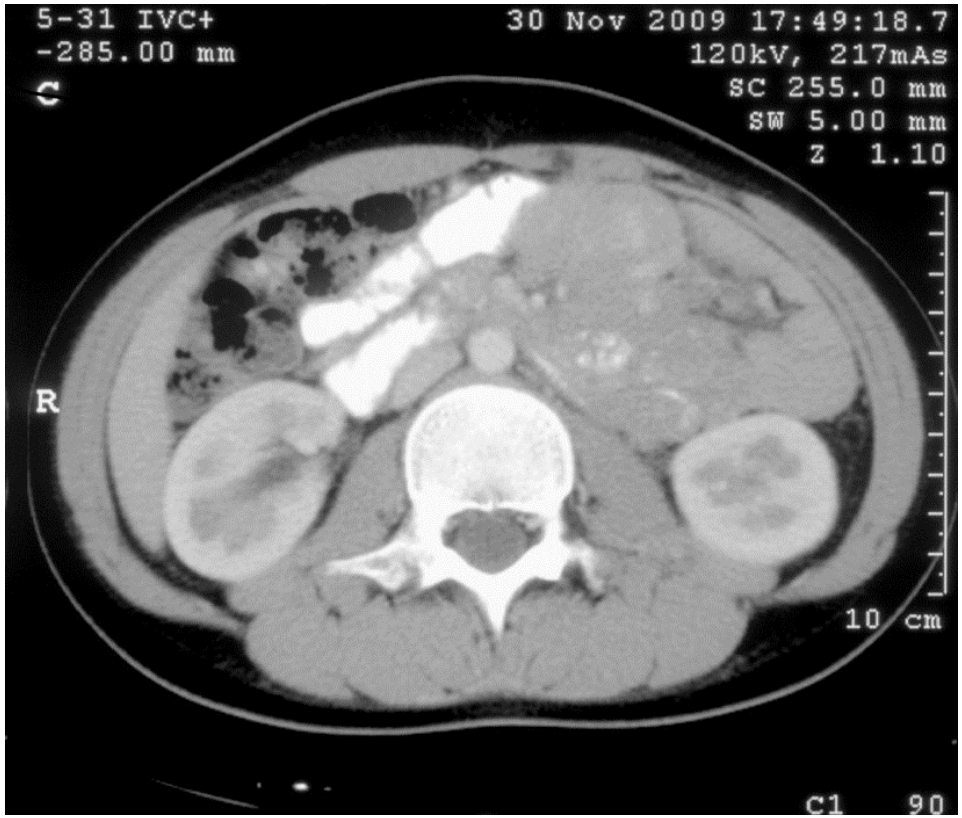
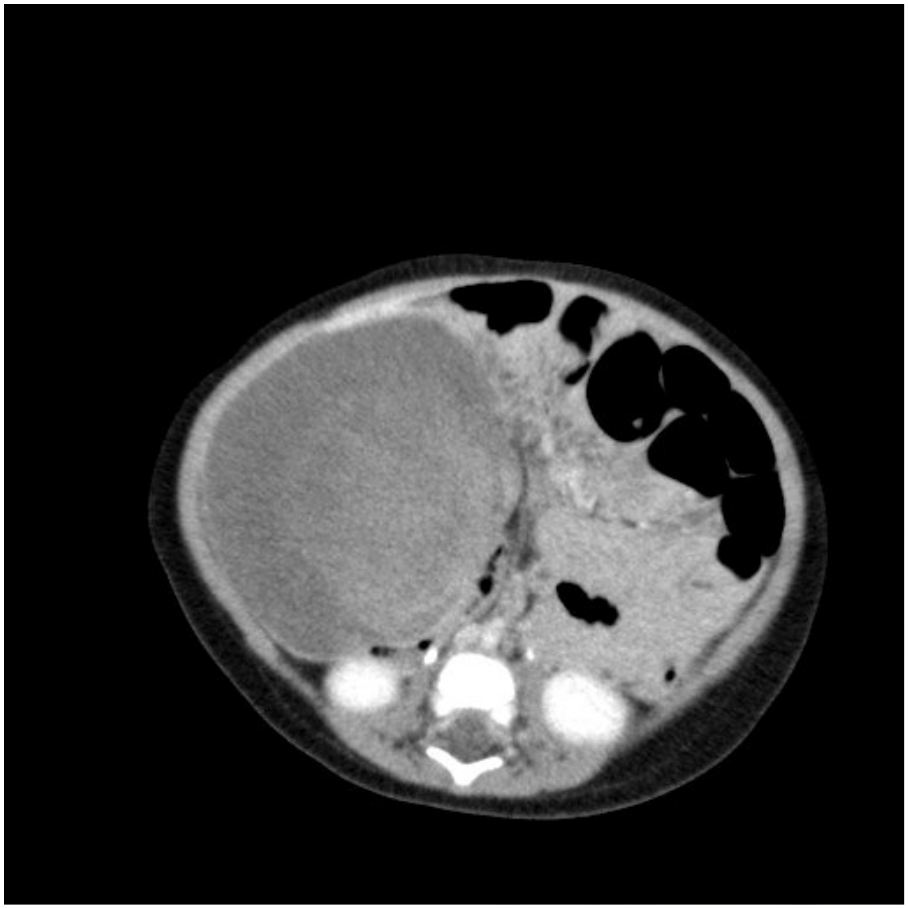
Discussion

Calcifying fibrous tumor (Case5)

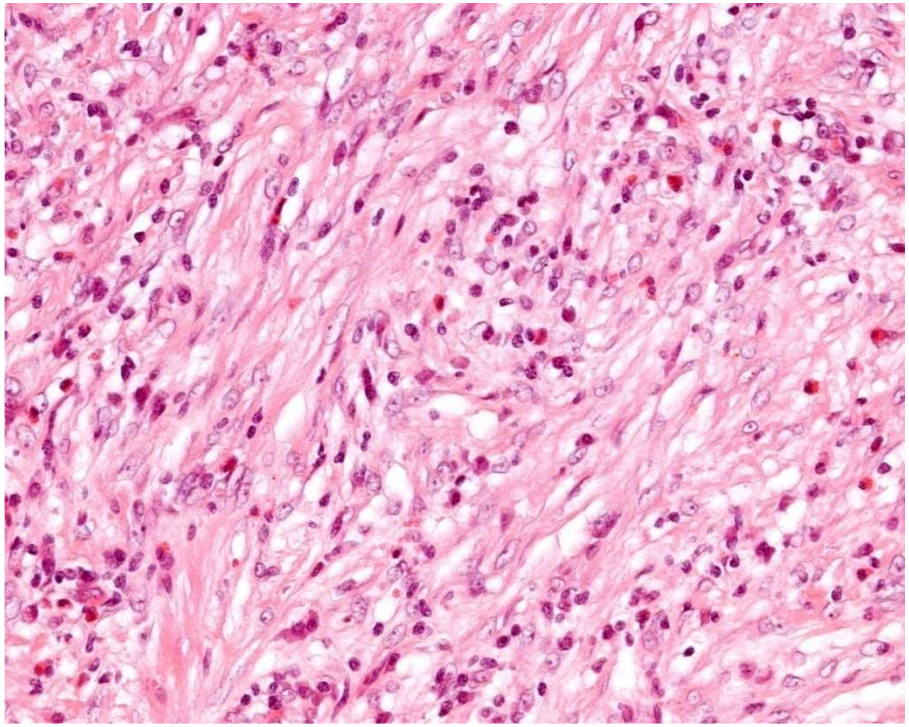


IMT (Case 4)

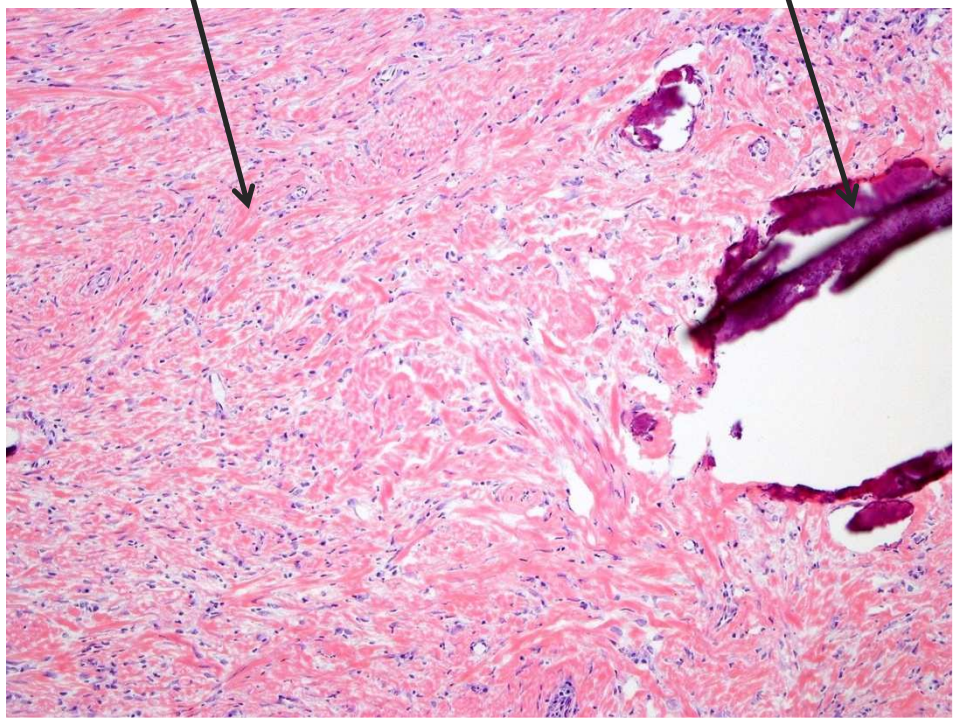


Discussion

Inflammatory myofibroblastic tumor

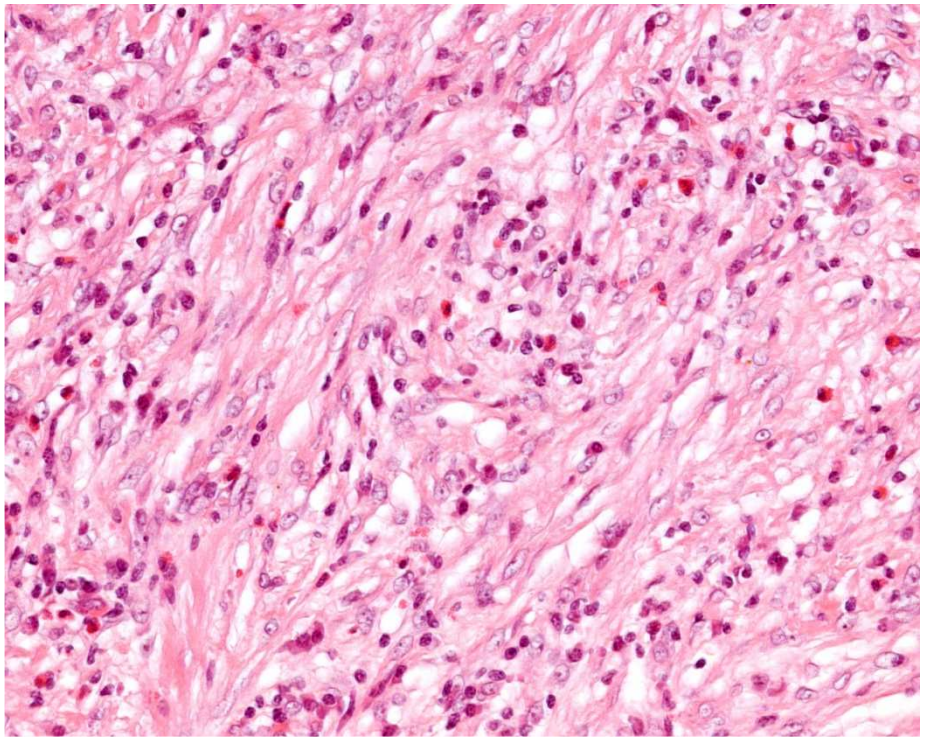


Calcifying fibrous tumor (Case 5)

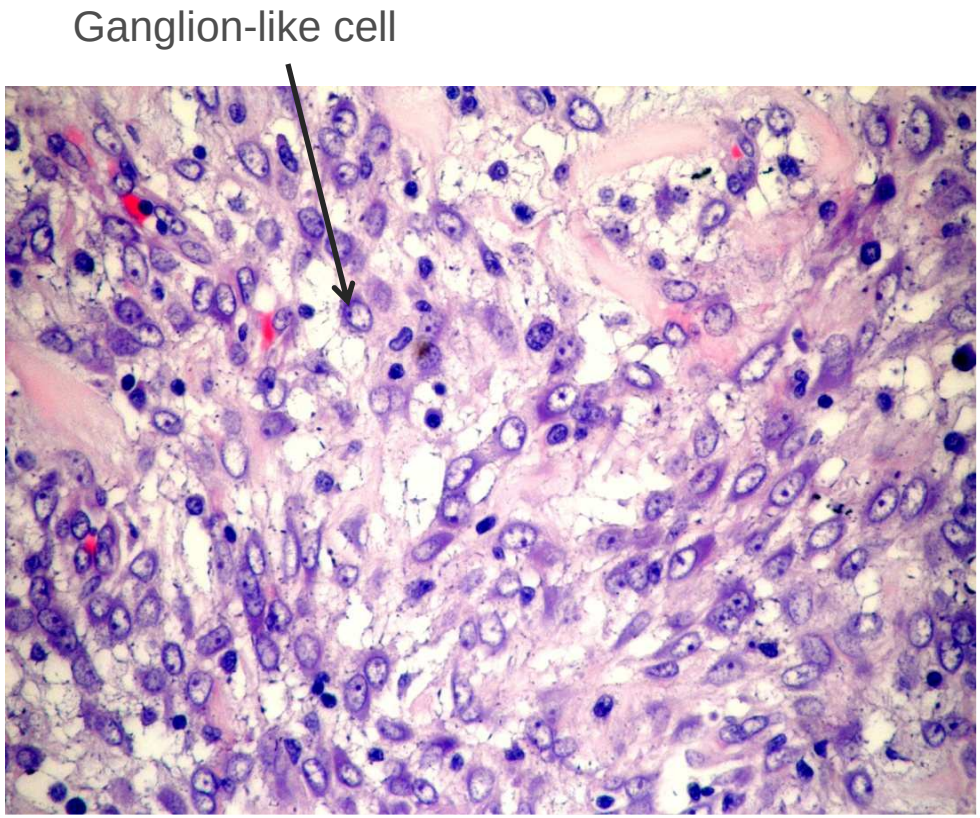


Discussion

Inflammatory myofibroblastic tumor



Inflammatory myofibroblastic tumor with ganglion-like cells (case 4)



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Conclusion

- Diagnostic difficulties
- Surgical resection as upfront management
- ALK gene mutations and ganglion-like cells: less favorable prognosis
- Calcifying pseudotumors do better
- Treatment difficulties in unresectable tumors

Thank You!

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