Pseudotumours In Mild Haemophiliacs – A Rare Pathology

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Background

- Pseudo tumours are chronic encapsulated enlarging haematomas caused by recurrent haemorrhage
- Availablity of clotting factor concentrates (CFC) in developed countries with established treatment programmes should prevent pseudotumours.
- Although rare (1-2%), they continue to cause disastrous complications in the developed world.

Objectives

1. To establish risk factors for pseudotumour formation in order to minimise later complications.
2. To demonstrate the applicability of MRI in differentiating pseudotumours from sarcomas

Methods

- Our tertiary referral centre is one of the largest in the UK.
- We performed a retrospective study of our database of haemophiliac patients over 10 years (2001-2011)
- Clinical presentation, management and complications of pseudotumours were analysed.

Results

- A total of 6 patients with possible pseudotumours were identified.(of which 2 were sarcomas)
- All patients had mild haemophilia A.
- Complications (resulting in lengthy ITU stay) included:
  - bowel obstruction
  - neurovascular compromise
  - bone erosion
  - renal failure
  - respiratory failure

CT - Large right pelvic pseudotumour causing iliac bone destruction

Patient	Age (yrs) at presentation	Factor VIII level (%)	Anatomical location	Pathology
1	26	7	Abdomen	Pseudotumour
2	15	6	Gluteals	Pseudotumour
3	57	8	Pelvis	Pseudotumour
4	31	12	Finger	Pseudotumour
5	74	6	Retroperitoneum	Sarcoma
6	36	38	Thigh	Sarcoma
Mean± SD	40±22	13±13		

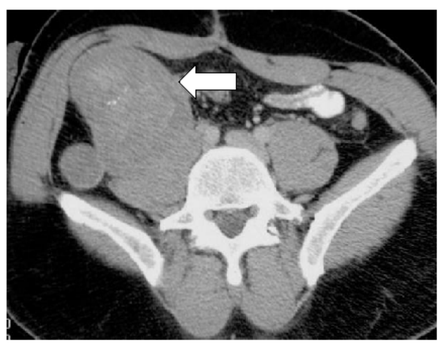
Essential Radiological Features

MRI is the best imaging modality for differentiating pseudotumours from sarcomas.

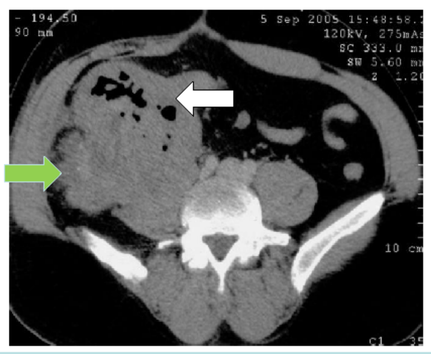
MRI	Pseudotumour	Sarcoma
Haemorrhage / Haemosiderin	++	Rare
Solid Components (excl haematoma)	Nil	+

Patient 1
- Increase size of RIF pseudotumour (over 13 years)
- Pseudotumour and colonic fistula.
- Evacuation of infected haematoma and right hemicolectomy
- Ongoing bleeding ⇨ 2nd laparotomy + angiographic embolisation
- Died aged 52

CT - large RIF pseudotumour compressing psoas muscle and common iliac vein

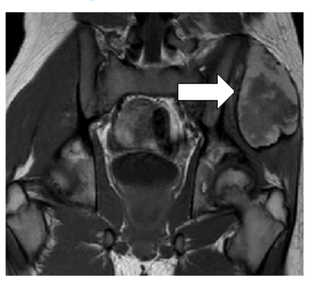


CT 1 year later - Fistula between pseudotumour (white) and colon (green)

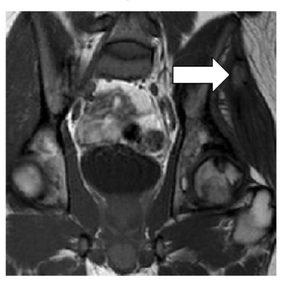


Patient 2
- Kicked in left buttock
- Initial inadequate treatment (lack of follow up)
- Persistent pain ⇨ MRI
- Pseudotumour diagnosed
- 3 months CFC
- Resolution of pseudotumour

MRI - Large pseudotumour within gluteal muscles

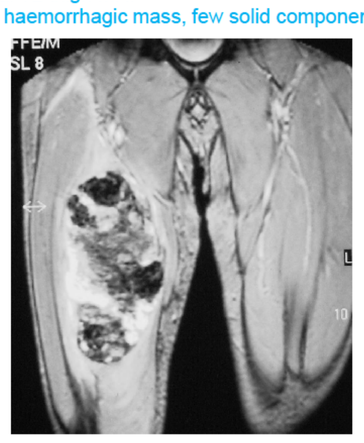


MRI 8 months later Resolution of pseudotumour



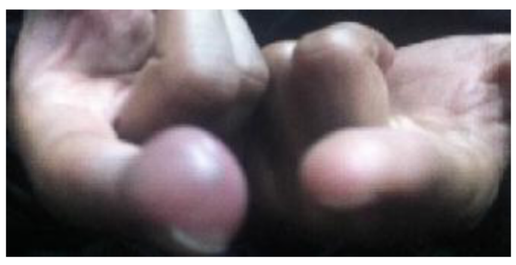
Patient 6
- Painful mass anterior thigh
- "Haematoma" diagnosed on MRI and at surgical exploration
- Biopsy of wall : SARCOMA
- Staging CT : lung metastases
- Died aged 36

Sagittal T2 thigh MRI – Large haemorrhagic mass, few solid components

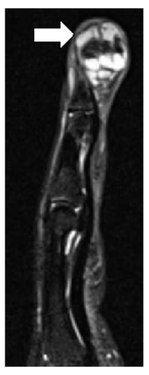


Patient 4
- Dropped 15kg weight onto tip of finger
- Only seen twice in 4 months following diagnosis and given FVIII
- Develops a 1 x 1.5cm pseudotumour
- Adequate CFC and surgical excision - resolution

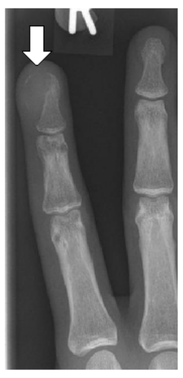
Right index finger mass



Sagittal MRI - pseudotumour causing periosteal elevation (arrow) and bone erosion



Xray showing bone erosion



Conclusion

- Mild/ moderate haemophilia patients continue to be a challenge for clinicians
- Present with disastrous complications in developed countries despite adequate supply of CFC as they:
  - Tend to ignore their bleeding problems and present late
  - Unable to self treat
  - Fail to attend for regular follow up
- Muscle haematomas require longer period of CFC treatment to prevent pseudotumour formation[1]
- MRI is the gold standard imaging modality

References
1. Sørensen B, Benson GM, Bladen M et al. Management of muscle haematomas in patients with severe haemophilia in an evidence-poor world. *Haemophilia* 2011 (in press)

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