Synovial Lipomatosis with Chondroblastoma in an 8-year-old Female: A Previously Unreported Entity

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Learning Point of the Article:
Timely diagnosis of synovial lipomatosis ensures appropriate management and decreased joint morbidity.

Abstract

Introduction: Synovial lipomatosis is a rare disease entity and a very small number of cases have been reported so far. It is characterized by villous proliferation of the synovium with expansion by mature adipose tissue. The etiology is unclear, thought cases can be seen secondary to injury, inflammation, chronic degenerative changes and neoplasms. Etiopathogenesis is still unclear, however is seen secondary to injury, inflammation, chronic degenerative conditions and neoplasms.

Case Report: An 8-year-old female child presented with pain and swelling in the left knee. Radiological examination suggested a lytic lesion in the upper tibia along with reactive synovial thickening. The lytic lesion was excised and an incisional biopsy was taken from the hyperplastic synovium. Histopathological examination of the synovial tissue showed villi-like structures with mature adipose tissue expanding the synovial lining along with the presence of mild chronic inflammatory cell infiltrate. The lytic lesion showed a cartilaginous tumor comprising mineralized chicken wire matrix surrounding the chondroblasts. A final diagnosis of synovial lipomatosis with chondroblastoma was made on histopathological examination.

Conclusion: This may be the first case report in medical literature of synovial lipomatosis coexisting with chondroblastoma in an adolescent girl. It also highlights the need for its increased awareness among young radiologists and pathologists so that an early diagnosis directs correct management and prevents further joint morbidity.

Keywords: Childhood, Chondroblastoma, Lipoma, Lipomatosis, Synovium.

Introduction

Synovial lipomatosis is an uncommon intra-articular proliferative lesion of mature adipose tissue causing villous transformation of synovium [1]. Chondroblastoma is a rare benign cartilaginous tumor arising from epiphyseal plates and apophyses of long bones such as humerus, femur, and tibia [2]. Extensive literature search failed to yield any case of chondroblastoma in association with synovial lipomatosis and this case may be the first case reported in medical literature. Both synovial lipomatosis and chondroblastoma are more commonly seen in males. Further, synovial lipomatosis is seen commonly in the 4th–6th decade of life. This case is rare not only due to its rarity but also due to its presentation in a pediatric age group female.

Case Report

An 8-year-old girl presented to the orthopedics OPD with swelling and pain in the left knee for the past 6 months (Fig. 1a). The swelling had rapidly increased in size in the past 1 month. There was a history of difficult in walking and frequent locking of this knee. No history of trauma or any other joint involvement could be elicited. No similar history was found in siblings or...
mother. An X-ray of the knee joint was advised which showed a lytic lesion in the proximal tibial epiphysis with peripherally placed sclerotic areas (Fig. 2a). X-ray did not reveal any significant joint effusion and in view of marked joint swelling, a magnetic resonance imaging (MRI) of knee joint was done. The MRI showed a heterogeneous lesion in the tibial epiphysis with altered signal intensity and a sclerotic rim (Fig. 2b). The lesion involved the tibial spine with a breach of the articular margin. There was evidence of perilesional edema with soft tissue enhancement suggesting a reactive pathology. On the basis of the above two, a differential diagnosis of tuberculosis and chondroblastoma was suggested. The hematological and biochemical work-up of the patient was within normal limits. The joint aspirate was negative for any crystals. Mantoux test read 8 mm. In view of no evidence of tubercular etiology and provisionally neoplastic radiological diagnosis, the patient was taken up for excision of the lesion with bone grafting. The bone graft was taken from fibula. Intraoperatively, the synovium appeared markedly hyperplastic, yellowish in color, and extending into the supra and infrapatellar compartments of the knee (Fig. 1b). The lytic lesion was managed by intralesional curettage and packing with allograft bone chips along with excision of the hyperplastic synovium. The curetted material from the lytic lesion and synovial tissue were sent for histopathological examination. Sections from the synovial tissue showed villi-like proliferations of the hyperplastic synovium with expansion of the subsynovium with mature adipose tissue (Fig. 3a and b) and also seen were interspersed chronic inflammatory cells and few dilated vessels. Sections from the curetted tumor material showed a cartilaginous tumor composed of fragments of mature hyaline cartilage, compact polyhedral chondroblasts with well-defined cytoplasmic borders and hyperlobulated nuclei with grooves. A chicken-wire matrix surrounding the chondroblasts was seen (Fig. 3c and d). No nuclear pleomorphism or mitosis identified. A final diagnosis of chondroblastoma with synovial lipomatosis was given. The MRIs were reviewed and it was seen that the soft tissue enhancement was in real the thickened synovium thrown in finger-like projections. Postoperatively, the pain and joint swelling improved drastically and no recurrence is seen till date.

**Discussion**

Synovial lipomatosis is a rare benign proliferation of mature adipose tissue cells in the synovial membrane. The most common site involved is suprapatellar pouch of knee joint. It is also known as lipoma arborescens and diffuse articular lipomatosis [3]. In 1904, this entity was first reported by Albert Hoffa, hence, the name Hoffa’s disease. Later on, the more elaborate term villous lipomatous proliferation of the synovial membrane as it is not a true neoplasm [3]. It commonly affects the older adults with median age of 50 years and very rarely does
Synovial lipomatosis

- Large frond-like appearance
- Secondary to chronic inflammation
- Usually painless
- Radiologically, signal intensity similar to fat

Characteristics

| Characteristic Features | Synovial lipomatosis | Synovial lipoma | PVNS | Synovial chondromatosis/chondrometaplasia | Oligoparticular JRA/RA |
|-------------------------|----------------------|-----------------|------|------------------------------------------|-----------------------|
| Etiology                | Unknown              | Unknown         | Unknown | Unknown                                  | Unknown               |
| Gross appearance        | Usually painless     | Occasionally    | Common | Sometimes                                 | Rare                  |
| Symptoms                | May show hemorrhage and hemosiderin | Yes | Yes | Yes | Yes |
| Clinical features       | Painless swelling    | Occasionally    | Usually | Sometimes                                 | Rare                  |
| Laboratory finding      | Normal               | Normal          | Normal | Normal                                   | Normal               |

Table 1: Characteristic features of various differential diagnoses

- **ETIOLOGY**
  - Synovial lipomatosis: Secondary to chronic inflammation.
  - Synovial lipoma: Secondary to trauma/inflammation.
  - PVNS: Secondary to trauma/inflammation.
  - Synovial chondromatosis/chondrometaplasia: Represents late stages of post-traumatic or degenerative lesions.
  - Oligoparticular JRA/RA: Unknown.

- **LABORATORY FINDING**
  - Synovial lipomatosis: Usually painless.
  - Synovial lipoma: Occasionally.
  - PVNS: Common.
  - Synovial chondromatosis/chondrometaplasia: Sometimes.
  - Oligoparticular JRA/RA: Rare.

- **RADIOLoGICAL FEATURES**
  - Synovial lipomatosis: Signal intensity similar to fat.
  - Synovial lipoma: Localized nodular mass.
  - PVNS: MRI – not usually done.
  - Synovial chondromatosis/chondrometaplasia: MRI – not usually done.
  - Oligoparticular JRA/RA: X-ray.

- **GROSS APPEARANCE OF SYNOVium**
  - Synovial lipomatosis: Large frond-like appearance.
  - Synovial lipoma: Small, solitary, polypoidal mass.
  - PVNS: Usually painless.
  - Synovial chondromatosis/chondrometaplasia: Numerous round osteocartilaginous nodules covering a thickened synovial surface.
  - Oligoparticular JRA/RA: Thickened and nodular synovium.

- **SYNOVIAL Lining hypertrophy**
  - Synovial lipomatosis: May show hemorrhage and hemosiderin.
  - Synovial lipoma: Atypical, binucleated forms.
  - PVNS: Usually painless.
  - Synovial chondromatosis/chondrometaplasia: Rare.
  - Oligoparticular JRA/RA: Rare.

- **VILLous transformation of synovium**
  - Synovial lipomatosis: Present.
  - Synovial lipoma: Absent.
  - PVNS: Present.
  - Synovial chondromatosis/chondrometaplasia: Present.
  - Oligoparticular JRA/RA: Present.

- **Chronic inflammation**
  - Synovial lipomatosis: Present.
  - Synovial lipoma: Absent.
  - PVNS: Absent.
  - Synovial chondromatosis/chondrometaplasia: Absent.
  - Oligoparticular JRA/RA: Absent.

- **TREATMENT**
  - Synovial lipomatosis: Excision through arthroscopic or open approach.
  - Synovial lipoma: Excision through arthroscopic or open approach depending on size.
  - PVNS: Surgical excision.
  - Synovial chondromatosis/chondrometaplasia: Complete surgical excision, 5% malignancy risk.
  - Oligoparticular JRA/RA: NSAIDs, corticosteroids, DMARDs.

- **BIOCHEMICAL FEATURES**
  - Synovial lipomatosis: Ablation with laser, embolization, and sclerosing agents.
  - Synovial lipoma: Ablation with laser, embolization, and sclerosing agents.
  - PVNS: Ablation with laser, embolization, and sclerosing agents.
  - Synovial chondromatosis/chondrometaplasia: Ablation with laser, embolization, and sclerosing agents.
  - Oligoparticular JRA/RA: Ablation with laser, embolization, and sclerosing agents.

This literature also describes its occurrence secondary to trauma, inflammation, degenerative joint conditions, and tumor’s emphasizing its metaplastic nature.

The clinical features of synovial lipomatosis include swelling in the affected joint, moderate intensity pain, and ipsilateral joint effusion. The joint effusion usually results from proliferation of synoviocytes. Pain in the joint may be due to the pressure effect of proliferating synovium or due to eroded articular cartilage secondary to any primary joint disorder. When knee joint is involved, it can even cause locking, joint line tenderness, and debilitation. Our patient also presented with swelling, pain, and frequent locking of the knee. The biochemical and culture analysis of the joint aspirate in such cases usually reveals no abnormality. X-ray is not diagnostic and the lesion may be...
missed, especially when focal and small in size. Ultrasonography shows a wavelike motion in cases of lipomatosis associated with intra-articular effusion [6]. MRI is the radiological investigation of choice for differentiating synovial lipomatosis from its closer mimics [5, 7]. Synovial lipomatosis characteristically shows fatty villous synovial proliferation without other signal intensities within the lesion on MRI [7]. Microscopically, it is characterized by diffuse involvement of the synovium with replacement by mature fat cells; however, sometimes, it can present as a solitary pseudomass [4, 7]. Another important characteristic feature is the presence of chronic mononuclear inflammatory cell infiltrate in the synovial tissue, indicating its origin secondary to chronic inflammatory conditions [4]. The present case also showed the presence of inflammatory cells along with fatty replacement of synovium.

The most common radiological and histopathological differential for synovial lipomatosis is synovial or intra-articular lipoma [6]. The common site of involvement for both is knee joint but can also involve hip, lumbar spine, and tarsometatarsal joint [8]. Clinical, radiological, and histopathological examination of the excised tissue has paramount importance in differentiating synovial lipomatosis from its other very close mimics, which is highlighted in Table 1 [1, 3, 4, 7, 8]. Asymptomatic cases need no intervention; however, symptomatic synovial lipomatosis can be temporarily treated with intra-articular corticosteroids, and the definitive treatment in such cases is arthroscopic excision of the synovium. Patients with extensive synovium involvement require open synovectomy [3]. This case report also intends to highlight the importance of effective clinical, radiological, and histopathological communication to reach to a diagnosis which not only affects patient treatment but also its prognosis.

**Conclusion**

This article aims at increasing the awareness of this entity among radiologists and orthopedic surgeons that although synovial lipomatosis is a very rare intra-articular lesion, it should be considered as differential diagnosis in any patient with any age presenting with a chronic soft boggy joint swelling. When diagnosed and treated on time, it will decrease the morbidity and joint abnormalities.

**Clinical Message**

Synovial lipomatosis is a rare entity but can be seen coexistent to non-neoplastic and neoplastic lesions of bones, contributing to the morbidity of the disease. MRI is the gold standard investigation for establishing the diagnosis. Early detection on radiology ensures timely intervention and reduced future joint abnormalities.

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