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Case Report

Massive Hemangiosynovial Tumor in a Child: A Case Report

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Abstract

Hemangiosynovial sarcoma is a rare vascular tumor that often presents in the joint synovium. This case report details a 13-year-old girl who presented with persistent knee pain, leading to the discovery of a mass on MRI, which was later diagnosed as hemangiosynovial sarcoma. The tumor, one of the largest reported in the literature, was evaluated with imaging techniques and treated successfully. A comparison with existing literature is provided, including a discussion of the incidence rate, diagnostic methods, and treatment options.

Keywords: *Hemangiosynovial Tumor; Magnetic Resonance Imaging (MRI); Pigmented Villonodular Synovitis (PVNS)*

Introduction

Hemangiosynovial, which is also called synovial hemangioma, is an uncommon benign vascular tumor that mostly affects the joint's synovial tissue. It is most frequently found in the knee joint, particularly in young people, and usually manifests as joint discomfort, swelling, and recurrent hemarthrosis. Because hemangiosynovial is so rare, it is challenging to estimate the total incidence; however, studies suggest that in the general population, the condition occurs 1 in 100,000 to 1 in 1,000,000 times [1,2]. Since most pediatric instances are detected in adolescence or early adulthood, the frequency is significantly lower in these circumstances. Less than 0.5% of all pediatric soft tissue tumors are thought to be pediatric hemangiosynovial, according to reports [3].

Given the rarity of hemangiosynovial in children, this tumor presents a notable diagnostic challenge. This case report details a rare presentation of hemangiosynovial in a 13-year-old girl, exploring its imaging and histological characteristics. Additionally, the management strategies employed in this case are discussed and compared with existing literature to provide a broader understanding of its clinical course and therapeutic outcomes in pediatric patients.

Case Report

A 13-year-old girl presented with chronic knee pain of insidious onset, without a history of trauma or systemic symptoms. The pain was primarily localized to the anterior aspect of the right knee and was exacerbated by physical activity. There was mild swelling, but no redness or fever.

An MRI of the knee was requested to further investigate the cause of the pain. The imaging revealed a well-circumscribed intra-articular mass, measuring approximately 3 cm in diameter, located in the suprapatellar region. The mass showed characteristic features of a hemangiosynovial tumor, including a lobulated appearance and areas of intermediate signal intensity on T1-weighted images, with marked hyperintensity on T2-weighted images. Post-contrast sequences demonstrated intense and homogeneous enhancement, consistent with the vascular nature of the lesion.

A biopsy was performed, and histopathological examination confirmed the diagnosis of hemangiosynovial tumor. The lesion was composed of proliferative small vascular channels lined by endothelial cells, consistent with a benign vascular tumor. No signs of malignancy were observed.

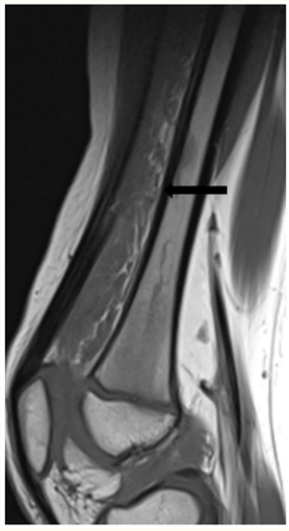

Figure 1: *Sagittal section T1 sequence without fat saturation showing a large mass in T1 isosignal at the level of the sub-quadricipital recess (arrows).*

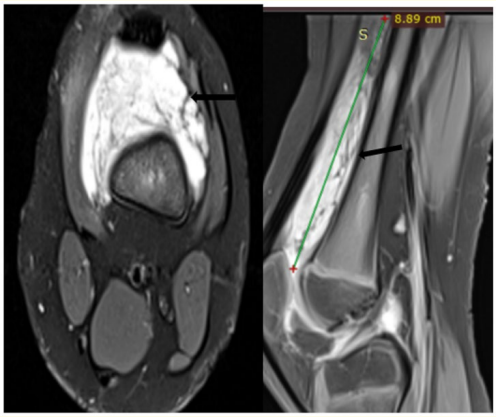

Figure 2: *Axial and sagittal section T2-weighted image with fat suppression technique shows the tumour with a high signal intensity, large-diameter measuring 8,89cm, thin fibrofatty septae of low signal intensity within the lesion (black arrow)*

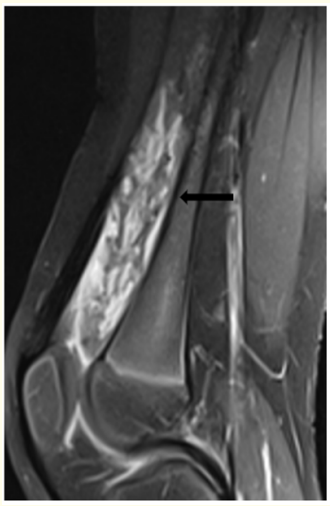

Figure 3: *Axial T1-weighted image after gadolinium administration demonstrates a mass of intermediate signal intensity with inhomogeneous enhancement in the suprapatellar pouch.*

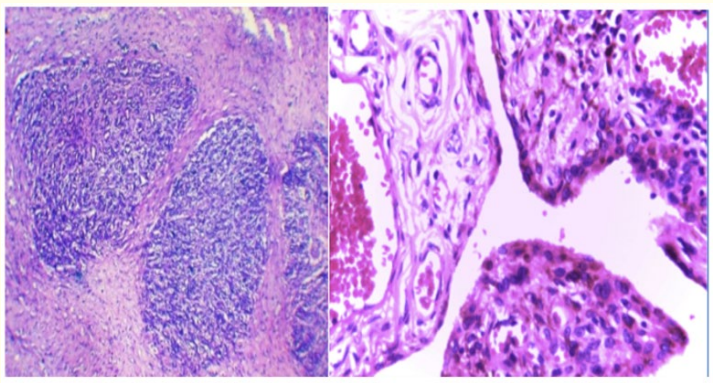

Figure 4: *The biopsy of hemangiosynovial tumors typically reveals proliferating blood vessels within the synovial membrane with a proliferation of capillaries surrounded by an important siderophagic reaction.*

Discussion

Hemangiosynovial tumors are uncommon, they are most frequently misdiagnosed leading to a diagnostic delay of many years; there are even reports of delays of up to 20 and 40 years, with a female predominance [4,5]. The knee joint is the most affected site, accounting for up to 70% of reported cases [6].

The clinical presentation of hemangiosynovial tumors is often nonspecific, and patients typically report pain, swelling, and limited range of motion in the affected joint. In children, these tumors often present with recurrent hemarthrosis, which may lead to a misdiagnosis of conditions such as juvenile idiopathic arthritis or hemophilia [7,8]. In our case, the absence of recurrent hemarthrosis, combined

with nonspecific symptoms, made the initial diagnosis more challenging, reinforcing the importance of thorough imaging and clinical evaluation.

The gold standard for diagnosing hemangiosynovial has consistently been demonstrated to be magnetic resonance imaging (MRI). Here, MRI showed a well-defined intra-articular mass with varying signal intensities, in line with findings published in the literature [9]. The mass showed hyperintensity on T2-weighted images and enhancement after contrast administration, indicating the high vascularity of the tumor. These imaging features are characteristic of hemangiosynovial and aid in distinguishing them from other joint-related pathologies like pigmented villonodular synovitis (PVNS) or synovial sarcoma [10].

In our case, the hemangiosynovial tumor measured 9 cm in its largest diameter, making it significantly larger than most cases reported in the literature. Typically, synovial hemangiomas range between 2 and 6 cm in size, as described by Xu., *et al*. (2018) [11] and Koudstaal., *et al.* (2007) who reported tumors of 3.5 to 6 cm in pediatric patients [12]. Larger tumors, such as the one described by Kumar., *et al*. (2014) [13] measured around 4 cm, while the study by Murphey., *et al.* (2001) [6] indicated sizes from 1.5 to 5 cm. Our case, with a 9 cm mass, highlights an unusually large presentation of this rare tumor, which likely contributed to the more pronounced clinical symptoms and the diagnostic challenge it posed. This size may also have increased the risk of complications, such as joint effusion and mechanical dysfunction, compared to smaller tumors.

Histologically, hemangiosynovial consist of proliferating blood vessels within the synovial membrane, forming thin-walled vessels arranged in small lobules. In this case, the biopsy confirmed the diagnosis by revealing these characteristic features. A similar histological pattern has been observed in other pediatric cases, reinforcing the critical role of biopsy in establishing a definitive diagnosis. This aligns with findings in pediatric synovial hemangiomas, where histopathology remains the gold standard for accurate identification and differentiation from other joint-related lesions [15].

Treatment of hemangiosynovial typically involves surgical resection, either through open or arthroscopic synovectomy. The decision for surgical intervention is based on the size of the lesion and the extent of symptoms. In our patient, arthroscopic synovectomy was successfully performed, with no recurrence of symptoms at follow-up. This outcome aligns with findings in the literature, where complete surgical resection is associated with a low recurrence rate. For instance, Moon., *et al*. (2014) reported a series of eight cases in which all patients underwent complete resection with no recurrence during follow-up [16]. Similarly, a study by Koudstaal., *et al*. (2007) emphasized that early intervention with complete resection leads to favorable outcomes, reducing the risk of complications and recurrence [17].

Conclusion

This case highlights the importance of considering hemangiosynovial tumors in the differential diagnosis of unexplained knee pain in pediatric patients. Early diagnosis through imaging and biopsy can lead to successful treatment, with complete surgical excision offering an excellent prognosis. Comparing this case with the literature underscores the rarity of the condition and the effectiveness of MRI in guiding diagnosis and treatment planning.

Further studies are needed to better understand the underlying pathogenesis of hemangiosynovial tumors and to explore potential non-surgical treatment options. However, current data suggest that surgery remains the gold standard for managing these lesions. Most reported cases have shown favorable outcomes following complete excision, with low recurrence rates and a good prognosis for long-term joint function.

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