Section: General Surgery

Case Report

LARGE ISOLATED VULVAL NEUROFIBROMA IN A CHILD: A CASE REPORT

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ABSTRACT

Background: Vulval tumors in the pediatric population are uncommon and often pose diagnostic and therapeutic challenges. Among these, neurofibromas are rare benign tumors of peripheral nerve sheath origin, typically associated with Neurofibromatosis Type 1 (NF1). Isolated vulval neurofibromas without systemic involvement are particularly unusual in children. Due to their rarity, large size at presentation, and potential for recurrence, these lesions require careful clinical evaluation, imaging, histopathological confirmation, and appropriate surgical management. This case report aims to highlight the clinical presentation, diagnostic approach, surgical management, and outcome of a large isolated vulval neurofibroma in a child. It also emphasizes the importance of considering rare benign tumors in differential diagnoses of vulval masses and discusses management challenges in pediatric patients. **Materials and Methods:** A 9-year-old female presented with a recurrent vulval swelling. Clinical examination, ultrasonography (USG), and magnetic resonance imaging (MRI) were performed to assess the lesion. An incision biopsy was conducted to establish histopathological diagnosis. The patient underwent wide local excision under general anesthesia, followed by histopathological evaluation of the excised specimen. **Result:** The patient presented with a large lobulated vulval mass measuring approximately 10×8 cm, involving the right labia majora with extension into adjacent thigh structures. Imaging revealed a multilobulated soft tissue mass with deeper extension into surrounding musculature. Biopsy confirmed myxoid neurofibroma. Complete surgical excision, including involved tissue planes, was performed successfully. Histopathology showed spindle-shaped cells with elongated nuclei consistent with neurofibroma. The postoperative period was uneventful, with preserved neuromuscular function and no immediate complications. **Conclusion:** Large isolated vulval neurofibromas in children are rare but should be considered in the differential diagnosis of vulval masses. Early diagnosis and complete surgical excision are crucial to prevent recurrence and complications. Multimodal evaluation and careful surgical planning are essential for optimal outcomes.

INTRODUCTION

Vulval tumors represent a heterogeneous group of lesions that may arise from epithelial, mesenchymal, or neural tissues. In the pediatric population, such tumors are exceedingly rare and often pose diagnostic challenges due to their varied presentation and overlapping clinical features. These lesions can broadly be categorized into cystic and solid tumors, with solid tumors including a wide spectrum of benign and malignant conditions. Among the benign mesenchymal tumors, neurofibromas are particularly uncommon in the vulval region.[1]

Neurofibromas are benign peripheral nerve sheath tumors composed of Schwann cells, fibroblasts, and perineural cells embedded in a myxoid stroma. They are commonly associated with Neurofibromatosis Type 1 (NF1), an autosomal dominant genetic disorder characterized by multiple neurofibromas, café-au-lait spots, and systemic involvement. Approximately half of vulval neurofibromas occur in association with NF1, while isolated cases without systemic features are rare. The occurrence of a large isolated vulval neurofibroma in a child is especially unusual and scarcely reported in medical literature.[2]

Clinically, vulval neurofibromas may present as slow-growing, painless masses. However,

depending on their size and location, they can cause discomfort, cosmetic concerns, and functional impairment. Large lesions may extend into adjacent structures, complicating surgical management. In children, such presentations necessitate careful evaluation to exclude malignancy and other serious conditions.[3]

The differential diagnosis of vulval masses is broad and includes benign lesions such as lipoma, papilloma, leiomyoma, lymphangioma, and angiomyxoma, as well as malignant tumors like sarcomas and carcinomas. Infectious and inflammatory conditions may also mimic tumor-like growths.

Therefore, a systematic diagnostic approach involving clinical examination, imaging, and histopathological evaluation is essential.[4]

Imaging modalities such as ultrasonography and magnetic resonance imaging (MRI) play a crucial role in assessing the extent, nature, and involvement of surrounding structures. MRI, in particular, provides superior soft tissue resolution and helps in surgical planning.[5]

Histopathological examination remains the gold standard for definitive diagnosis, typically revealing spindle-shaped cells with elongated nuclei and a myxoid background in neurofibromas.[6]

Management of vulval neurofibromas primarily involves surgical excision. Complete removal of the tumor is essential to prevent recurrence. In cases of large or infiltrative lesions, excision may involve adjacent tissues and require reconstructive procedures using local flaps or grafts. Preservation of function and cosmetic outcomes are important considerations, particularly in pediatric patients.[7]

This case report describes a rare presentation of a large isolated vulval neurofibroma in a 9-year-old girl with a history of recurrence. The case highlights the importance of early diagnosis, appropriate imaging, and complete surgical excision. It also underscores the need to consider rare benign tumors in the differential diagnosis of vulval swellings in children.[8]

CASE DESCRIPTION

A 9-year-old female presented to the outpatient department with a complaint of recurrent swelling over the right side of the vulva for one year. The swelling was insidious in onset and gradually increased in size over time.

Figure 1: Pre-operative examination

There was no history of pain, fever, trauma, discharge, bleeding, paraesthesia, or skin discoloration. The patient did not report any similar swellings elsewhere in the body.

There were no visual disturbances or neurological symptoms. Her past medical history was significant for a prior surgical excision of a similar swelling at the same site in 2023. There was no relevant family history or associated comorbidities.

On local examination, a large lobulated swelling measuring approximately 10×8 cm was noted on the right side of the vulva. The mass involved the right labia majora, extended laterally into the right thigh, and overhung towards the left labia majora. The surface was nodular with well-defined but irregular margins. The swelling was firm to hard in consistency and had limited mobility. The overlying skin was adherent at the center, corresponding to the previous surgical scar, and free at the periphery.

Ultrasonography revealed multiple lobulated hypoechoic soft tissue lesions that appeared coalescent, suggestive of a benign soft tissue tumor. Magnetic resonance imaging (MRI) of the pelvis demonstrated deeper extension of the lesion into the right adductor longus, gracilis muscle, and pubic symphysis. An incision biopsy was performed, and histopathological examination indicated features consistent with myxoid neurofibroma.

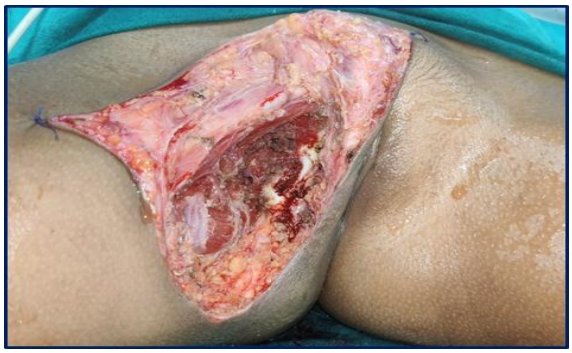

Figure 2: Wound bed after excision of swelling

The patient underwent wide local excision of the tumor under general anesthesia. The central adherent portion of the skin, including the previous surgical scar, was excised along with the tumor. Portions of the gracilis muscle and tissue adherent to the pubic bone were also removed due to tumor

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involvement. The defect was reconstructed using local skin flaps, and a suction drain was placed.

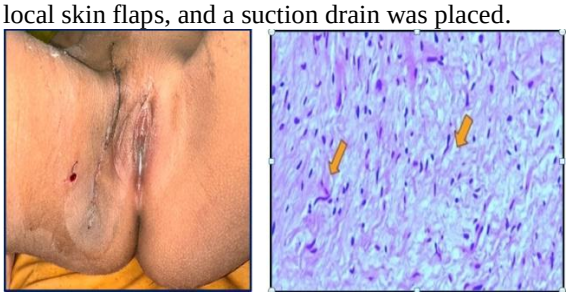

Figure 3: (A) Drain was removed on post-op day 3, (B) Histopathology: Spindle cells having narrow elongated nuclei with tapered ends and indistinct cytoplasm

Postoperatively, the patient recovered well with intact neuromuscular function. The drain was removed on postoperative day three, and the patient was discharged in stable condition. Histopathological examination of the excised specimen confirmed the diagnosis of neurofibroma, showing spindle-shaped cells with elongated nuclei and indistinct cytoplasm.

DISCUSSION

Neurofibromas are benign tumors arising from the peripheral nerve sheath and are commonly associated with Neurofibromatosis Type 1 (NF1). However, isolated neurofibromas without systemic manifestations are rare, particularly in the vulval region and even more so in pediatric patients. The present case represents a rare occurrence of a large, recurrent, isolated vulval neurofibroma in a child, highlighted important diagnostic and therapeutic considerations.[9]

Vulval neurofibromas constitute a small percentage of benign vulval tumors, estimated at around 5%. Their rarity often leads to delayed diagnosis or misdiagnosis. Clinically, they may present as painless, slow-growing masses, but larger lesions can cause discomfort, functional impairment, and cosmetic concerns. In this case, the tumor reached a considerable size and showed local extension into adjacent musculature, emphasizing its infiltrative potential.[10]

The differential diagnosis of vulval masses is extensive and includes benign tumors such as lipoma, leiomyoma, papilloma, schwannoma, rhabdomyoma, and angiomyxoma, as well as conditions like lymphangioma, hamartoma, and angioedema. Malignant lesions must also be considered, particularly in rapidly growing or recurrent cases. Therefore, accurate diagnosis relies on a combination of clinical evaluation, imaging, and histopathology.[11]

Imaging plays a crucial role in assessing the extent of the lesion. Ultrasonography is often the first-line modality, providing information about the nature of the mass. MRI is particularly valuable in delineating soft tissue involvement and guiding surgical planning. In this case, MRI revealed infiltration into the adductor muscles and proximity to the pubic symphysis, which influenced the surgical approach.[12]

Histopathological examination remains the definitive diagnostic tool. Neurofibromas are characterized by spindle-shaped cells with elongated nuclei, set in a myxoid stroma. The absence of atypia and mitotic activity helps distinguish them from malignant peripheral nerve sheath tumors.[13]

Surgical excision is the treatment of choice for vulval neurofibromas. Complete removal is essential to minimize the risk of recurrence. However, achieving clear margins can be challenging in large or infiltrative lesions, particularly in anatomically complex regions like the vulva. In this patient, wide local excision with removal of involved adjacent structures was necessary.[14]

Reconstruction of the surgical defect is an important consideration. Depending on the size and location of the defect, local flaps, grafts, or more complex reconstructive techniques may be required. In this case, local skin flap adjustment was sufficient to achieve satisfactory closure.[15]

The recurrence observed in this patient may be attributed to incomplete excision during the initial surgery. This underscored the importance of thorough surgical planning and complete tumor removal. Casabona F, et. al; 2010, highlighted the need for a high index of suspicion, comprehensive evaluation, and meticulous surgical management in rare vulval tumors in children.[16]

Clinical Significance: This case highlights the importance of considering rare entities such as isolated vulval neurofibroma in the differential diagnosis of pediatric vulval masses. Early recognition and appropriate evaluation are essential to avoid misdiagnosis and delayed treatment. The case emphasizes the role of imaging, particularly MRI, in determining the extent of the lesion and guiding surgical planning. Recurrent lesions should raise suspicion of incomplete prior excision, necessitating a more aggressive surgical approach. Complete excision with adequate margins is critical to prevent recurrence and achieve optimal outcomes. Additionally, careful intraoperative management is required when the tumor involves adjacent structures. The absence of systemic features of Neurofibromatosis Type 1 in this patient underscores the need to differentiate isolated neurofibromas from syndromic cases. Reporting such rare cases contributes to medical literature and enhances clinician awareness, ultimately improving patient care and outcomes in similar presentations.

CONCLUSION

Large isolated vulval neurofibroma in children is a rare clinical entity that requires a high index of suspicion for diagnosis. Comprehensive evaluation using imaging and histopathology is essential to confirm the diagnosis and assess tumor extent. Surgical excision remains the cornerstone of

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treatment, with complete removal being crucial to prevent recurrence. This case demonstrates that even large and recurrent lesions can be successfully managed with appropriate surgical planning. Long-term follow-up is recommended to monitor for recurrence. Increased awareness of such rare presentations will aid in early diagnosis and improved management of pediatric vulval tumors.

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