Lipoblastoma in infant: Our experience

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Lipoblastoma is a rare benign soft tissue tumor that occurs most commonly in infants and children. The wide majority are detected in children under the age of 3 years. Lipoblastoma usually present as a rapidly growing mass most often located in the trunk and extremities. Lipoblastoma has rarely been reported in the inguino-labial region. No malignant degeneration has been documented while a high recurrence rate has been described and is usually correlated to the diffuse-type lesions (lipoblastomatosis) and to incomplete excision.

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Lipoblastoma, first described by Jaffe [1] in 1926, is an uncommon benign soft tissue tumor, affecting infancy and early childhood and consisting of lipoblasts that continue to proliferate during the postnatal period. The wide majority are detected in children under the age of 3 years with 80/90% of cases occurring before the age of 3 and 40% before the age of 1 [2]. There is a male predominance of 3:1. About 70% of these tumors occur in the extremities, trunk, head and neck. Lipoblastomatous lesions contain a spectrum of fat cells ranging from immature lipoblasts to mature adipocytes. We report two cases of lipoblastoma in infant and discuss surgical management and recurrence.

1. Methods

A 12-month-old white female infant presented with a history of a progressively enlarging left labial mass. There was no history of trauma or infection. Physical examination revealed the labia major entirely occupied by a mass that encompassed her left labium majus and perineal region with characteristics suggestive of adipose tissue. Surgical excision via a left labia incision approach demonstrated a well-encapsulated, soft, yellowish-white mass that was completely excised (Fig. 1). Microscopic examination the tumor was composed of mature fat and myxoid changes with lipoblasts among the adipocytes. Three years follow-up has not revealed any signs or symptoms of recurrence.

A six-months white female infant presented at the Paediatric Surgery Department for a painless superficial swelling in the left inguinal region. Her parents reported that the mass had appeared one month before. She underwent surgery. In the subcutaneous tissue there was a non-capsulated mass with macroscopic appearance of a yellowish tissue (2×1 cm), and a total gross excision was believed to be achieved. At histology it was defined as a lipoblastoma.

Three months later the patient presented with an enlarging subcutaneous hard mass of 7×5×6 cm, rapidly increasing of size, above the initial incision and extended to the labia majora and inguino-crural left region interpreted as local recurrence (Fig. 2). Preoperative abdominal US scan was done to identify other potential sites of lipoblastomatosis. Through a mini suprapubic left extended incision the mass was completely removed. Post-operatively a lymphatic subcutaneous collection was aspirated three times. At present there is no evidence of local recurrence or lymphatic collection. Pathology confirmed the preoperative diagnosis of recurrent lipoblastoma; the mass was almost totally circumscribed with fibrous connective tissue.

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2. Discussion

Lipoblastoma is a relatively rare tumor that occurs in infancy and early childhood and arises from embryonic white fat. Histopathologic examination shows a cellular neoplasm composed of immature adipocytes with relatively well-defined septa, frequent lipoblasts and a fine vascular network. The most important clinical aspect in accurate recognition of lipoblastoma and lipoblastomatosis lies in their distinction from liposarcoma, especially its myxoid variant. According to Mentzel et al. [3] liposarcoma is rare in patients under 10 years and in myxoid form, it may be almost impossible to distinguish it histologically from lipoblastoma. Helpful clues are the lack of lobulation variable growth pattern, and increased nuclear atypia in liposarcoma. Mahour et al. [4] suggested that lipoblastoma would eventually differentiate into a mature lipoma.

Lipoblastoma, the focal circumscribed form, tends to occur superficially and macroscopically it seems to be a lipoma while lipoblastomatosis, the diffuse and infiltrative form, tends to occur in deeper tissue and has a higher recurrence rate [5]. Clinically the most common symptom is a painless mass with or without increasing size. The trunk, extremities, head and neck, retroperitoneum, peritoneal cavity, and lung are the common tumor sites. Rarely, as in case 1, lipoblastoma has been described in the inguino-labial region [6].

3. Conclusion

Despite its potential to invade locally and a grow rapidly to a large size, this tumor has an excellent prognosis. Metastases have never been described. It is now well established that lipoblastoma best treatment is complete surgical excision to avoid leaving residual tumor and to prevent recurrences, which mostly occurs within 2 years. A regular and periodic follow up is necessary for many years because the recurrence rate in local or marginal excision range from 12 to 25% in spite of complete excision [3].

References