DERMATOFIBROSARCOMA PROTUBERANS IN A 13-YEAR-OLD: A RARE CASE OF SOFT TISSUE SARCOMA OF THE SKIN

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue sarcoma that primarily affects middle-aged adults but can also occur in children. Despite its low metastatic potential, DFSP displays aggressive local behavior and a high rate of local recurrence. Diagnosis in children can be delayed due to confusion with other skin conditions. This case report aims to discuss the case management of dermatofibrosarcoma protuberans in a 13 year old female.[1,2]

CASE REPORT

Report A 13-year-old female patient presented with a progressively enlarging lump on the left eyelid, initially resembling small brown spots that coalesced over 7 months. Intermittent bleeding and pain were reported

PHYSICAL EXAMINATION

Physical examination revealed a dark brown mass in the left frontotemporal region, with solid, non-fragile characteristics with indistinct borders, an uneven surface, and fixed dimensions measuring 3x2x1 cm.

MANAGEMENT

The management of patient were wide excision and flap reconstruction. Wide excision aim for removal of the dark brown mass from the left frontotemporal region. Flap reconstruction performed to ensure closure and aesthetic appearance. The procedure was completed without any intraoperative complications. Follow-up imaging and clinical assessments at subsequent visits showed no signs of recurrence or additional complications with only dry wounds that felt painful and itchy on day 6 after surgery.

HISTOPATHOLOGICAL FINDINGS

Histopathological examination results concluded pigmented dermatofibrosarcoma protuberans in the left frontotemporal region, and the surgical wound margins did not contain malignant tumor cells.

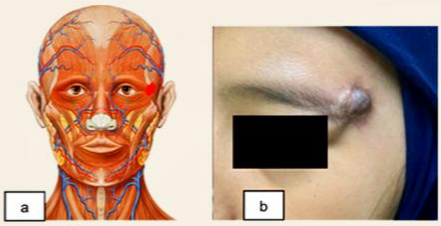

Figure 1. (A) The position of the lesion on frontotemporal sinistra region (B) Clinical examination of DFSP

Figure 2. Post operative (H+1) examination

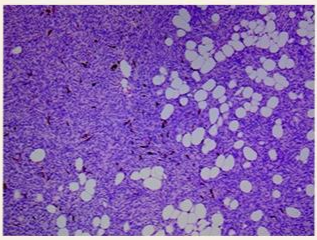

Figure 3. Histological features

DISCUSSION

DFSP in recent studies indicate an equal distribution between genders and manifest in the neonatal period or at any later stage, with an estimated incidence of approximately one per million in individuals under 20 years old. Early-stage DFSP can be mistaken for other skin conditions like hypertrophic scars, keloids, cysts, and lipomas. As DFSP progresses, it can grow outward, become thin, hard, or ulcerated, and even spread to deeper tissues. In later stages, DFSP may resemble other cancers like melanoma, Kaposi sarcoma, and other soft tissue sarcomas. Surgical excision is the primary modality for the treatment of DFSP, aimed at achieving complete tumor resection. Mohs micrographic surgery allows for precise removal of the tumor. Histopathological examination is indispensable to verify the completeness of tumor removal. Microscopic examination reveals a unique growth pattern of DFSP cells, extending from the skin into the adipose tissue. Conventional chemotherapy often doesn't work well for DFSP. Radiation therapy is often used after surgery. Regular follow-up after radiation therapy is important. Regular monitoring aims to improve patient outcomes by detecting recurrences early and allowing for prompt therapeutic intervention.[3,4]

CONCLUSION

(DFSP) is a type of soft tissue cancer that affects young to middle-aged adults. The primary treatment is surgery, with wide local excision being the typical approach. For smaller lesions in sensitive areas, Mohs micrographic surgery may be considered. Radiation therapy and targeted therapy can be used as alternative or additional treatments. Regular follow-up is crucial for early detection of recurrence or metastasis.

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