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연소성 황색육아종에서 피부 로사이-도프만병으로의 이행

(Cutaneous Rosai-Dorfman Disease Following Juvenile Xanthogranuloma)

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Purpose: Rosai-Dorfman disease (RDD) is a rare histiocytic disorder. While the skin is the most common extranodal site, purely cutaneous RDD (CRDD) is exceptionally rare, accounting for only 3% of cases. Both xanthogranuloma and RDD are non-Langerhans cell histiocytoses, often posing diagnostic challenges. We report a rare case where an initial Juvenile xanthogranuloma(JXG) transitioned into CRDD, suggesting a potential phenotype change

Methods: A 12-year-old girl presented with a 7 x 6 mm skin mass on her left cheek. To minimize scarring, a staged excision was planned. Partial excision was initially performed, with histopathology suggesting JXG. Fifteen months later, a complete excision was performed. The specimen underwent comprehensive immunohistochemical staining. Systemic involvement was assessed via contrast-enhanced CT and laboratory evaluations.

Results: The first-stage histopathology suggested xanthogranuloma with S100-negativity. However, the second-stage specimen revealed pathognomonic features of RDD, such as emperipolesis, and showed positivity for CD68, S100, and Cyclin D1, while testing negative for CD1a, Langerin, and OCT2. Systemic evaluation confirmed the absence of lymphadenopathy or extranodal involvement, diagnosing purely CRDD. Genetic analysis for the MAPK-ERK pathway mutations showed no known mutations, including BRAF. Postoperatively, the wound healed well, and the patient is undergoing scar management with no recurrence or systemic symptoms

Conclusion: This case illustrates a rare histological evolution from JXG to CRDD over 15 months within the histiocytic lineage. Clinicians should be alert to such potential transformations; longitudinal monitoring and repeat biopsies in evolving lesions are essential for accurate diagnosis and appropriate systemic management.

Fig. 1A. Preoperative photograph for the first-stage excision. To minimize the final scar length, a partial excision was strategically planned as the initial step.

Fig. 1B. Preoperative photograph for the second-stage excision performed 15 months later.

Fig. 1C. Clinical follow-up at one month postoperatively, demonstrating stable wound healing without any complications and a minimized scar following the staged-excision approach.