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Nevus Sebaceous: A Midline Verrucous Lesion on a Neonate

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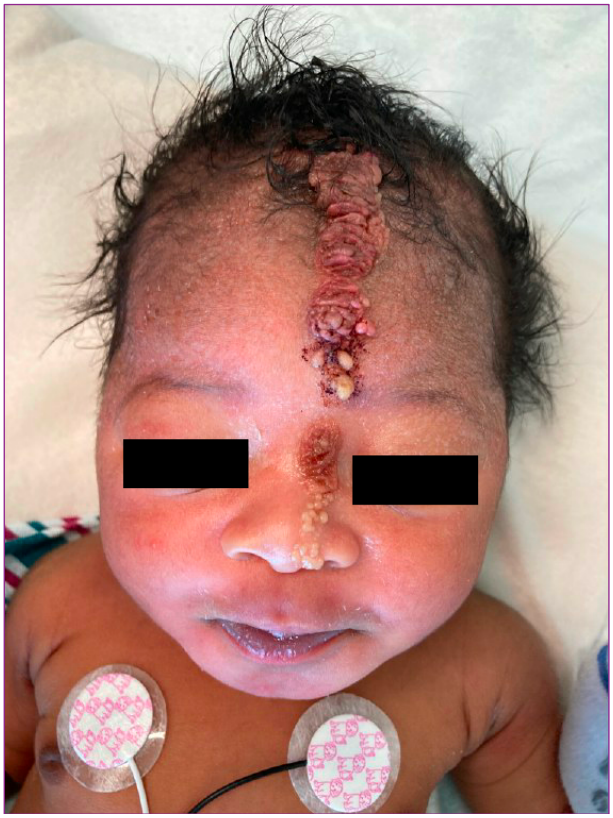

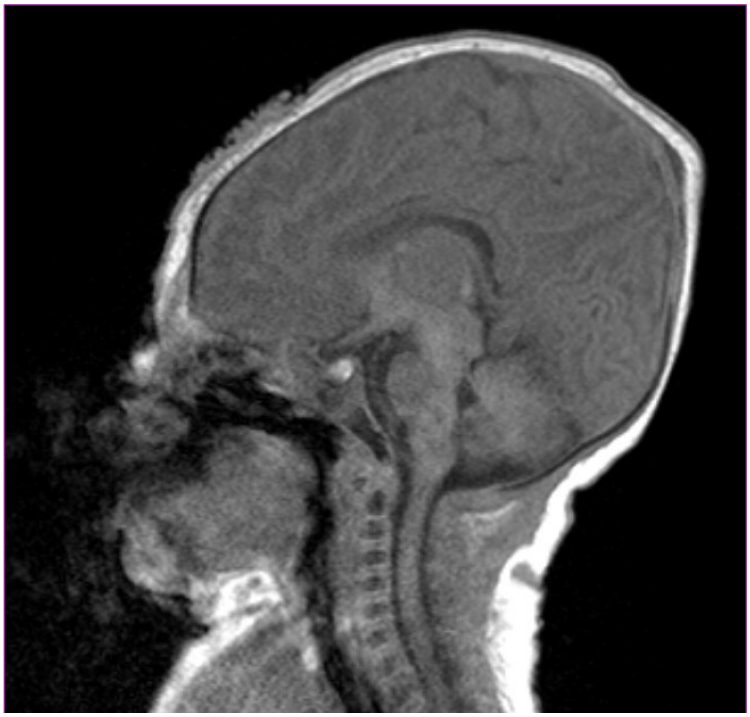

CASE REPORT

Dermatology was consulted for a newborn girl with a midline verrucous lesion extending from the forehead to the nasal tip. The infant received little prenatal care, but no abnormalities were noted on 11 or 20-week ultrasound. The teenage G2P1 mother was taking Keppra during pregnancy and delivered at 39 weeks via low transverse caesarian section. MRI of the brain, x-ray of the spine, and neurology and ophthalmology consults were recommended to rule out skeletal, central nervous system (CNS), or ocular involvement. Ophthalmology found no ocular complications. MR imaging showed a normal brain and exophytic lesions without intracranial communication. Chest and vertebral x-ray were unremarkable. Patient

met developmental milestones at 1-week and 2-month pediatric visits.

DISCUSSION

Nevus sebaceus is a congenital malformation of the pilosebaceous follicular unit thought to be caused by postzygotic mosaic mutations in the HRAS and KRAS genes. It affects approximately 0.3% of newborns with an equal incidence across sex and race. Lesions occur most commonly on the scalp as an oval or linear, smooth, yellow, well-circumscribed plaque. Patients with large or disseminated lesions can have underlying CNS or skeletal abnormalities.[1]

Diagnosis of nevus sebaceous is often clinical. Histopathologic features of the lesion in infancy include immature pilosebaceous units with acanthosis and mild papillomatosis. As the lesion grows, the sebaceous glands become more prominent, and an increasing number of malformed ducts are seen.

Nevus sebaceus can be associated with several clinical syndromes. Schimmelpenning syndrome, or nevus sebaceous syndrome, is caused by an autosomal dominant, lethal mutation in HRAS or KRAS genes which survives due to somatic mosaicism.[2] The original triad of symptoms included the presence of a sebaceous nevus, seizures, and intellectual disability; however, the syndrome is now defined as the association of a linear nevus sebaceus with extracutaneous anomalies of the ocular, nervous, skeletal, and cardiovascular systems. Phacomatosis pigmentokeratotica is another associated syndrome that presents with a papular nevus spilus and linear nevus sebaceous, typically in addition to associated extracutaneous abnormalities.[3] These patients are at an increased risk for internal malignancies such as rhabdomyosarcoma and nephroblastoma in addition to basal cell carcinoma and melanoma.[4]

Other benign or malignant neoplasms may arise from a nevus sebaceous, and the malignancy risk increases with age. The lifetime risk of a malignant neoplasm arising from a nevus sebaceous is unknown. One study found out of 5,000 cases, 16% of patients developed benign lesions and 8% of patients developed malignant lesions. Basal cell carcinomas are the most common malignant neoplasm to arise within a nevus sebaceous, and it is possible for more than one lesion to exist in the same nevus.[5]

Management is somewhat controversial. Full thickness-excision is the definitive treatment; however, given the low risk of secondary malignancy in children, excision may be unnecessary. Alternative treatments include photodynamic therapy, carbon dioxide laser resurfacing, and dermabrasion. Simple, solitary lesions in children do not routinely require additional evaluation; however, with extensive lesions, a neurologic and ophthalmologic examination and imaging, such as MRI, may be required to rule out associated clinical syndromes.

Consent: The authors obtained informed consent from the patient's guardians prior to the write up and inclusion of patient images in this report

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