A localized scleroderma-like reaction in a giant congenital nevus

A. Patsatsi¹, V. Lambropoulos², I. Efstratiou³,
M. Kokolios¹, O. Pikou¹, D. Sotiriadis¹

1. 2nd Department of Dermatology, Aristotle University Faculty of Medicine, Papageorgiou General Hospital, Thessaloniki, Greece
2. Department of Pediatric Surgery, Aristotle University Faculty of Medicine, Papageorgiou General Hospital, Thessaloniki, Greece
3. Department of Pathology, Papageorgiou General Hospital, Thessaloniki, Greece
Case

- A 6 year old girl – one of dizygotic tweens after in vitro fertilization
- zosteriform plaque on the lower abdomen, pubic and lumbosacral area
- on palpation, the lesion was uniformly sclerotic
- foci of pigmentation, reminders of a giant congenital nevus
Photographic material from infancy was available for evaluation of the lesion's evolution during the past 6 years.
Photographic material from infancy was available for evaluation of the lesion’s evolution during the past 6 years.
The sclerodermiform reaction process seems to have started in utero, as at birth, there were also sclerotic areas and more pigmented foci that seemed to have regressed six years later.
Specimen from a heavily pigmented site
Specimen from a sclerotic area
Discussion

• This a rare presentation of a giant congenital nevus with a progressive sclerodermoid reaction, described in a few papers in the literature

• Patee SF. et al, in 2001, first described a giant congenital nevus (GCMN) with an unusual progressive “sclerodermoid” reaction in a newborn

• Two years later, Ruiz-Maldonado R. et al proposed the term ‘desmoplastic hairless hypopigmented nevus’ for this entity

• In 2007, Hernandez-Martin A. et al, proposed the term ‘sclerotic hypopigmented GCMN’, based on pathological grounds
Pathogenesis

- fibrotic stromal change may represent an atypical host reaction to nevus cells

- a multidirectional differentiation of melanocytes into adaptive fibroblasts and tumor extracellular matrix interactions may lead to the induction of collagen synthesis

- this reaction may start in utero and last as long as the immune system recognizes nevus antigens
Prognosis

- none of the reported cases (12, so far) has been found with melanoma during the follow up period

- extended excision should be avoided as it results in unnecessary disfigurement

- any malignant potential of sclerotic hypopigmented GCMN has to be elucidated throughout adulthood
References


