

Neonatal Cutaneous Myxoma

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Abstract: The association of myxoid subcutaneous tumors with pigmented skin lesions. Patients with rare skin and soft tissue lesions suffer from delays in diagnosis and subsequently care. It is difficult for the clinician to keep up with the most current treatment recommendation since literature review is very few. Our institution benefits from multi-specialty efforts as evidenced from the case aforementioned. It is recommended that multidisciplinary approach is necessary for optimal management.

[Sarah ElNassir ElNigoumi, Babiker Kambalwi Hassan. **Neonatal Cutaneous Myxoma.** *J Am Sci* 2017;13(4):60-62]. ISSN 1545-1003 (print); ISSN 2375-7264 (online). <http://www.jofamericanscience.org>. 7. doi:[10.7537/marsjas130417.07](https://doi.org/10.7537/marsjas130417.07).

Keywords: association; myxoid; subcutaneous tumor; skin lesion; patient; tissue

1. Introduction

The association of myxoid subcutaneous tumors with pigmented skin lesions and atrial myxoma was recorded for the first time by Atherton et al.(1), who used the acronym NAME (nevi, atrial myxoma, myxoid neurofibroma, endocrine overactivity) to designate this syndrome. Rhodes et al(2) in 1984 described a second patient with similar findings and renamed the syndrome LAMB(lentiginos, atrial myxoma, mucocutaneous myxoma, blue nevi) syndromes, after noticing that the pigmented lesions were lentiginos and blue nevi and the cutaneous tumors were myxomas. In 1985 Carney et al(3) discussed the association of spotty pigmentation with myxomas (cardiac, mammary and cutaneous) and endocrine disorders. The latter included primary pigmented nodular adrenocortical disease manifest by Cushing's syndrome, various testicular tumors and growth hormone-producing pituitary adenomas. Several other authors(4,5,6) have reported patients described as having Carney's complex.

2. Case Presentation

This is a newborn baby girl, who was born at term by spontaneous vaginal delivery, to an epileptic mother on Lamotrigine and positive history of group B hemolytic streptococci on HVS, with APGAR score of 9 and 9 at 1 & 5 minutes respectively, and birth weight of 3.19kg.

The neonate was noted to have an oval cystic swelling on the posterior aspect of the head and was kept in neonatal ICU for investigation and further management and was initially thought to have a meningocele which was not diagnosed antenatally.

The patient was admitted to the NICU and kept in incubator. In view of her swelling, initial impression of meningocele was made.

Physical examination revealed a small oval shaped, cystic swelling, measuring approximately 2x3 cm on the posterior fontanelle, which was non-tender, and immobile. The overlying skin did not show any evidence of fluctuation nor erythema.

Systemic examination was normal and did not reveal any other spinal cord or midline defects.

Furthermore, we observed no evidence of neurological deficit.

She was then reviewed by our neurosurgeons who decided to take the child for repair of the meningocele after conducting MRI of brain and spinal cord.

The brain MRI revealed the presence of an oval shaped cystic swelling measuring 22.8 x9.7 mm overlying the posterior fontanelle with no visible brain tissue within lesion.

A complete blood cell study was done along with a blood culture in view of maternal infection.

Complete blood cell showed a white cell count of $13.85 \times 10^9/L$; hemoglobin of 20.8g/dL and platelet count of $196 \times 10^9/L$. and blood culture was sterile for 120 hours.

Spine MRI was normal with no evidence of spina bifida or associated defects.

Meningocele repair was done by the neurosurgery team on April 7, 2016; with excisional biopsy of the suspected meningocele.

During the repair, it was found that the lesion did not contain CSF nor brain tissue, which made the suspicion of meningocele less likely.

Macroscopic examination of the scalp skin lesion showed a 25 x 2 x 1 cm cyst filled with grayish brown

material. Microscopic examination showed few strands of fibroblasts and proliferating vessels, with no evidence of malignancy, leading to the diagnosis of cutaneous myxoma.

3. Differential Diagnosis

Many skin lesions such as cutaneous myxoid fibroblastoma, focal cutaneous mucinosis and superficial angiomyxomas maybe considered when diagnosing, making the differences in diagnoses according to clinical appearance, histopathology, vascularity and epithelial components.



Cutaneous myxomas are flesh colored and mildly translucent and well circumscribed. Microscopically, they are made up of sparse spindle/stellate cells within the myxoid matrix with delicate reticulum and collagen fibers with no nuclear polymorphism and prominent capillaries making these

lesions the highest suspected differential diagnosis in this case.

Per literature review, this is the first published case documenting Neonatal Cutaneous Myxoma.



4. Discussions

Cutaneous myxomas without other components of the Lamb's syndrome or the Carney's complex is a very rare occurrence. In fact, a literature search revealed no similar cases. Ditz & Stout(7) in 1961 described 15 myxomas of the external soft tissues in children which did not originate in the skin properly, but in the subcutaneous tissue. Johnson & Helwig(8) in 1966 described 14 cases of cutaneous focal mucinosis in 14 patients. They were single lesions classified as focal mucinosis rather than as true myxomas. Other reports have described multiple lesions of papular mucuosis associated with thyroid abnormalities (9). Digital mucous cysts and ganglion cysts are also included in the category of muconosis and are not considered to be true myxomas. Single cases of cutaneous myxomas have also been described (10). Unlike other patients, all the previously published cases of multiple cutaneous myxomas have been associated with one or more of the other components of the Carney's complex which include:

- (i) single or multiple cardiac myxomas,
- (ii) single or multiple myxoid tumors at various cutaneous and mucosal sites,
- (iii) multiple lend genes/ freckles predominantly in the centofacial area, but also involving mucosal surfaces, particularly the conjunctiva, and blue nevi,
- (iv)myxoid mammary fibroadenomatosis,

(v) a variety of rare endocrine tumors including: Leydig cell and large-cell neuroendocrine tumours,

Johnson & Helwig described the histologic features of cutaneous myxomas which allow differentiation from cutaneous mucinoses. They included:

(i) an extensive network of reticular fibers in myxomas,

(ii) absence of collagen fibers in myxomas as opposed to focal mucinosis, and

(iii) more sharply delineated margins of myxomas(11).

Carney also described the histologic features of cutaneous myxomas including:

(i) either upper dermal, dermal and subcutaneous, or purely subcutaneous location,

(ii) sharp circumscription but without encapsulation in the dermis, and with occasional encapsulation in the subcutis, a prominent basophilic or acidophilic myxoid stroma containing proteoglycan,

(iii) Hypocellularity,

(iv) prominent vascularity with multiple regularly disposed capillaries,

(v) lobulation,

(vi) the presence at times of an epithelial component(12).

4. Conclusion:

Patients with rare skin and soft tissue lesions suffer from delays in diagnosis and subsequently care. It is difficult for the clinician to keep up with the most current treatment recommendation since literature review is very few.

Our institution benefits from multi-specialty efforts as evidenced from the case aforementioned.

It is recommended that multidisciplinary approach is necessary for optimal management.

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3/25/2017