Syringocystadenoma papilliferum
developed on sebaceous nevus. Three pediatric cases.

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Summary
Sebaceous nevus is a congenital organoid nevus. It is mainly located on the scalp and appears as a hairless yellowish plaque at birth or soon after. Sebaceous nevus changes with growth being sebaceous glands responsive to hormones at puberty. Since benign and malignant tumors can arise within sebaceous nevus, short follow-up and surgical excision are probably the better medical behavior to manage these lesions when changes appear. Here are reported three pediatric cases of syringocystadenoma papilliferum developed in the context of sebaceous nevus.

Key words
Sebaceous nevus, syringocystadenoma papilliferum.

Sebaceous nevus is a common congenital lesion occurring mainly on the head, particularly on the scalp. It was firstly described in 1895 from Jadassohn as a hamartomatous lesion predominantly composed of sebaceous glands.

Pinkus defined this disease as “organoid nevus” being the anomalies not only relative to sebaceous glands but also to sweat glands and hair follicles (8).

Sebaceous nevus appears as an alopecic yellowish patch at birth or early childhood and tends to become raised, papillomatous at puberty as a result of the effect of the androgen hormones on the sebaceous glands.

Numerous benign and malignant tumors are known to arise within sebaceous nevus (5). The commonest benign neoplasms are syringocystadenoma papilliferum (SCAP) and trichoblastoma, whereas the commonest malignant one is basal cell carcinoma (BCC). The incidence of these tumors increases with age, in particular after puberty.

Another significant association of sebaceous nevus is a neurocutaneous syndrome known as linear nevus sebaceous syndrome (Schimmelpenning-Feuerstein-Mims syndrome, Solomon syndrome), which is a subtype of the wider epidermal nevus syndrome. The commonest extracutaneous manifestations in patients with epidermal nevus syndrome involve the central nervous system, ocular and skeletal systems (5).

Here we report three cases of SCAP arising on sebaceous nevus in childhood and discuss the therapeutic approach to these complex lesions.

Case report

We present three cases of SCAP arising on sebaceous nevus in pediatric patients. The data of our patients are summarized in Table 1.
Table 1: Data of patients with syringocystadenoma papilliferum arisen on sebaceous nevus.

<table>
<thead>
<tr>
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<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
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<tbody>
<tr>
<td>Tumor</td>
<td>SCAP</td>
<td>SCAP</td>
<td>SCAP (Fig. 1)</td>
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<tr>
<td>Age (years)</td>
<td>12</td>
<td>10</td>
<td>15</td>
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<tr>
<td>Site of SN</td>
<td>Trunk</td>
<td>Left temporal area</td>
<td>Frontal (hairline)</td>
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<td>Clinical presentation</td>
<td>Nodule</td>
<td>Nodule (Fig. 2)</td>
<td>Nodule</td>
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<tr>
<td>Excision</td>
<td>Entire lesion</td>
<td>Nodule</td>
<td>Entire lesion</td>
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Discussion

Sebaceous nevus is an organoid nevus present mostly at birth or early childhood in 0.05-1% of newborns (1). It presents as an alopecic yellowish plaque mainly located on the scalp and face. Other localizations are rare but possible.

Three clinico-pathological stages have been identified in the evolution of this congenital lesion. In the first, infantile stage, clinically it appears with smooth surface and yellowish-orange color. Histopathologically it shows slightly acanthotic and hyperpigmented epidermis and in the dermis hair follicles are small and incompletely formed, often represented by solid cords of basoid, undifferentiated cells. Sebaceous glands are not prominent. In the second, adolescent stage, sebaceous nevus becomes clinically thicker with surface nodules or verrucous hyperkeratosis. Histologically, there is verrucous epidermal hyperplasia and in the dermis hyperplasia of sebaceous glands with small hair follicles, which remain primordial. Often also apocrine glands with dilated lumina and hyperplasia of eccrine glands can be found. The third, adult stage, is characterized by epidermal hyperplasia, large sebaceous glands and ectopic apocrine glands. The hair follicles remain primordial. In the last stage several benign and malignant tumors can occur (7).

Among the benign tumors arising on sebaceous nevus Shapiro et al. (7) include 1- tumors with pilar differentiation such as trichoblastoma, desmoplastic trichilemmoma, trichoepithelioma, follicular and pilar hamartoma, proliferating trichilemmal cyst, follicular infundibuloma; 2- basaloid neoplasms with follicular differentiation such as epidermal inverted follicular keratosis; 3- tumors with sebaceous differentiation such as sebaceous adenoma, sebaceous epithelioma; 4- tumors with apocrine differentiation such as tubular apocrine adenoma, apocrine cystadenoma, apocrine hidrocystoma, apocrine epithelio...
Syringocystadenoma papilliferum on sebaceous nevus

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Among the malignant tumors arising in sebaceous nevus the same Authors (7) include: basal cell carcinoma, squamous cell carcinoma, keratoacanthoma, apocrine carcinoma, eccrine porocarcinoma, sweat gland carcinoma, anaplastic salivary gland adenocarcinoma, sebaceous carcinoma, adnexal carcinoma with pilar differentiation and mucoepidermoid carcinoma.

Many works have been done to define the risk of development of tumors in the context of a sebaceous nevus. However, during the past years there was no agreement between the Authors. Only recently different large studies reached the same conclusions. Cribier et Al. (2) found a total of 13.6% of benign tumors -less than 2% in sebaceous nevus excised in children, 0% under 10 years- and 0.8% of malignant -none in children- in their 596 cases. Santibanez-Gallerani et Al. found no basal cell carcinoma (BCC) in their sebaceous nevus series of 658 cases of patients aged 16 years or less. They did not found either benign tumors (6). Kaddu et Al. analyzed 316 cases of SN and found 7.6% of benign tumors -only in adults- and two malignant tumors -only in adults as well- (4). Munoz-Perez et Al. found 18% of benign tumors in their series of 226 patients, all in subjects above the age of fourteen (5). These Authors agree about the conclusion that tumors arising on SN are exceptional under the age of ten years and almost rare until puberty. After this period the incidence increases with age.

In adulthood also malignant tumors can appear in until 40% of sebaceous nevus (1). SCAP and trichoblastoma are the most frequent benign tumors, while BCC is the most frequent malignant one. Although it is somewhat difficult to differentiate BCC from trichoblastoma arising in sebaceous nevus, some studies have outlined a number of morphologic criteria useful in differentiating these two neoplasms (4).

For a pediatric dermatologist the matter is to decide if a child with sebaceous nevus must be followed up during years or if he/she must be addressed to surgery. Probably the correct behavior is to inform the patient and/or his/her parents about the possible occurrence of this complication and its rate according to the age. When a tumor or bleeding occurs, the patient should contact the dermatologist. In case of suspected tumor, the latter will advise removal of the entire nevus or only the tumor, according to the individual case. After puberty the patient should be informed about the need for a dermatological examination in case of clinical changes of sebaceous nevus. In this event surgical excision of the lesion must be undertaken.

Indeed it is of importance to underline that, even if the most frequent tumor arising in sebaceous nevus is a benign tumor as SCAP, it can exceptionally evolve in its malignant counterpart, syringocystoadenocarcinoma papilliferum, with important consequences on the prognosis (3).

In conclusion, our 3 cases confirm the data of the literature regarding the age of risk for developing of tumors within congenital sebaceous nevus. Our youngest patient was 10-years-old and the other 2 patients were older. Also the histologic type of tumor of our patients is in accordance with the data reported from the literature, being SCAP the most frequent neoplasm which can arise in a sebaceous nevus, developing in until 19% of the of sebaceous nevus (7). Interestingly, two of our patient had a sebaceous nevus of the head, while the third had a sebaceous nevus of the trunk. The latter localization is quite peculiar because several Authors agree that most tumors in sebaceous nevus are usually located on the scalp (2).
References