A Naevus Sebaceous with Tumour of the Follicular Infundibulum, Trichilemmoma, Desmoplastic Trichilemmoma, Apocrine Adenoma and Syringocystadenoma Papilliferum: Report of a Case

Coyne JD1*, Chatzipantelis P2

1The Royal Oldham Hospital, Rochdale Road, Oldham OL1 2JH, United Kingdom
2Department of Pathology, Medical School, Democritus University of Thrace, Dragana Alexandropoulis, Greece

*Corresponding Author: Coyne JD, The Royal Oldham Hospital, Rochdale Road, Oldham OL1 2JH, United Kingdom; Email: johnnycoyne@doctors.org.uk

Received Date: 18-05-2020; Accepted Date: 26-05-2020; Published Date: 31-05-2020

Copyright© 2020 by Coyne JD, et al. All rights reserved. This is an open access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Nevus Sebaceous of Jadasson (SNJ) has been reported with various coexisting benign and rarely with malignant neoplasms. We present a case of SNJ with five benign epidermal and adnexal neoplasms, some of them with unusual features. Histologically, all of the typical findings of SNJ appear in our case along with a) a focal area of basaloid proliferation (tumour of the follicular infundibulum), b) more desmoplastic area with features of desmoplastic trichilemmoma and c) a nodular area with cytological features of trichilemmoma. In addition d) apocrine adenoma and e) syringocystadenoma papilliferum were present in adjacent areas. There were no features of malignancy. In summary, we report a complex case of SNJ with five unusual and concurrent neoplastic variants.

Keywords

Naevus Sebaceous; Apocrine Adenoma; Trichilemmoma; Desmoplastic Trichilemmoma; Basaloid Follicular Hamartoma; Syringocystadenoma Papilliferum
Introduction

Nevus Sebaceous of Jadassohn (SNJ) is a cutaneous hamartoma that has a well-documented potential to develop a variety of benign and, less commonly, malignant neoplasms of epidermal and adnexal origin [1]. Most common neoplasms are syringocystadenoma papilliferum, trichoblastoma and basal cell carcinoma [2,3]. Other reported neoplasms include spiradenoma, squamous cell carcinoma, sebaceous carcinoma, syringocystadenoma papilliferum, apocrine carcinoma, trichilemmoma and mucoepidermoid carcinoma [1,4-6]. We report a case of SNJ with multiple and unusual neoplastic components.

Case History and Pathology

A 59 year old woman had a long standing lesion, clinically diagnosed as a naevus sebaceous with suspected basal cell carcinomatous change on her posterior scalp. The excised specimen measured 7×5 mm. Microscopic examination showed verrucous, acanthotic, hyperplastic papillary surface proliferation with subjacent apocrine and sebaceous glands and separate hair follicles, features consistent with a nevus sebaceous (Fig. 1). In addition, a focal area of basaloid proliferation was present (Fig. 2). This resembled a basaloid follicular hamartoma (tumour of the follicular infundibulum) and merged with an adjacent area focally desmoplastic and focally nodular. No malignant features were identified. The lesion was completely excised. The first, desmoplastic area consisted of a dermal proliferation of bland polygonal cells with clear cytoplasm and arranged in small clusters and elongated strands, features consistent with a desmoplastic trichilemmoma (Fig. 3). Towards the deeper aspect of the tumour there were large lobules of similar bland cells with clear and glassy cytoplasm, peripheral palisading and an outer thickened eosinophilic basement membrane consistent with a trichilemmoma (Fig. 4). In addition, a focus of small tubular glands with eosinophilic cytoplasm were present towards the surface resembling an apocrine adenoma. Moreover, several separate papillary foci were present with an endo and exophytic papillary proliferation respectively lined by glandular cells with numerous plasma cells in the subjacent fibrous stroma consistent with a syringocystadenoma papilliferum (Fig. 5). A focus of dystrophic ossification was present.
**Figure 1:** Features of a naevus sebaceous.

**Figure 2:** Basaloid follicular hamartoma.

**Figure 3:** Desmoplastic trichilemmoma.

Coyne JD | Volume 1; Issue 1 (2020) | JDR-1(1)-006 | Short Communication


DOI: http://dx.doi.org/10.46889/JDR.2020.1201

DOI: http://dx.doi.org/10.46889/JDR.2020.1201

Figure 4: Trichilemmoma.

Figure 5: Syringocystadenoma papilliferum and apocrine adenoma.
Discussion

Multiple neoplasms may arise occasionally within SNJ, it is rare for four or more neoplasms to occur simultaneously [7]. Many benign and malignant tumours may arise within the postpubertal stage of this lesion [8]. The frequency of development of neoplasms is in direct proportion to the age of the patients [3]. Most of these neoplasms are follicular, apocrine or sebaceous tumours due to their common embryonic origin [2]. Our present case involves 5 neoplasms in a solitary lesion.

Neoplasms arising with SNJ were TFI, TL, DTL, AA and SCAP; TFI, TL and DTL are of follicular origin, while AA and SCAP are of apocrine gland origin. Trichoblastoma and SCAP are the most common neoplasms associated with SNJ [2]. Trichilemmoma and TFI are less common as the previous ones. Desmoplastic change surrounding the nodular area of TL is uncommon finding in this neoplasm [9]. Tumour of the follicular infundibulum poses differential diagnostic difficulties with other more common neoplasms such as basal cell carcinoma. Epidermal basaloid proliferation raised the possibility of a superficial basal cell carcinoma. However, retraction artefact and myxoid stroma were not present. Also, superficial trichoblastoma was included in the differential diagnosis. Many common histological features make a final diagnosis more difficult. Many neoplasms arising in SNJ do not correspond precisely to well describe entities and are difficult to classify histologically [10]. Apocrine adenoma and SCAP are tumours of apocrine gland origin. It is important to distinguish adenomas from well differentiated adenocarcinoma.

Due to the fact that the tumour in this case reported was small and without an infiltrative pattern of growth facilitated our diagnosis.

Nine previous reports have documented the occurrence of four or more neoplasms arising in a SNJ [1,7,11-16]. These have been summarised and tabulated by Dore et al. [16]. Four comprised of four neoplasms, two comprised of five neoplasms, two comprised of six neoplasms and one of seven tumours. All findings confirm the position of SCAP, trichoblastoma and TFI as the most common elements occurring in association with a SNJ. Our report adds one more rare complex case with multiple unusual neoplastic components.

Currently, prophylactic excision in SNJ is considered the optimal treatment [1,7]. Clinical features are not sufficient to make an exact diagnosis of secondary benign or malignant tumours. Therefore, excisional biopsy is recommended for better histological assessment and close clinical follow-up is also advised.

Reference


