Smyrna Tıp Dergisi ***Olgu Sunumu***

The Case of Fibrous Papule Developing on the Nose: A Case Report
Burunda Gelişen Fibröz Papül Olgusu: Olgu Sunumu

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Summary
Fibrous papules (also known as angiofibromas in several sources) are defined as clinically benign, asymptomatic, dome-shaped, skin-colored, generally single, small, and hard lesions. From the clinical aspect, they might be confused with the melanocytic nevus, wart, or small nodular type basal cell carcinoma (BCC). From the histological aspect, the fibrous papule consists of fibrous stroma containing generally widespread and floret-like multinucleated CD34-positive cells, fibroblast on the superficial dermis, dilated blood vessels, and fibrovascular proliferation. Many types of it such as classical hypercellular, clear cell, pigmented, epitheloid, pleomorphic, inflammatory, and granular types have been identified. The presented case was a 29-year-old woman who was referred to the plastic surgery polyclinic by her family doctor because of lesions on the chin and lateral side of the nose. The patient had no characteristic in her history other than the iron deficiency anemia and the polyp in gallbladder. In the physical examination, a off-white and brown-colored lesion on the chin was found to have 0.7cm diameter and the lesion on the nose was found to have off-white color, dome shape, and 0.3cm diameter. As a result of the histopathological analysis of lesions, the lesion on the chin was diagnosed with intradermal nevus, whereas the lesion on the nose was diagnosed with fibrous papule because of its morphological characteristics and Factor XIIIA weak (+) and CD34 (+) staining panel. The present case is presented here together with literature since fibrous papule might be confused with benign or malign skin lesions.
Key words: Fibrous papule, nose, young female patient

Özet
Fibröz papüller (ayrıca çeşitli kaynaklarda anjiyofibromlar olarak da bilinir) klinik olarak benign, asemptomatik, kubbe görünümlü, ten renkli, genellikle tek, küçük, sert lezyondur. Klinik olarak genellikle melonositik nevüs, siğil veya küçük nodüler tip BCC (bazal hücreli karsinom) ile karışabilirler. Histolojik olarak fibröz papül yüzeyel dermiste fibroblast, genişlemiş kan damarları, fibrovasküler proliferasyon, dağınık, yıldız şeklinde sıklıkla multinükleer CD 34 pozitif hücreler içeren fibröz stromadan oluşmaktadır. Klasik hiperselüler, berrak hücreli, pigmente, epiteloid, pleomorfik, inflamatuar ve granüler olmak üzere birçok tipi tanımlanmıştır. Sunulan olgu, çene ve burnun yan tarafındaki lezyonlar nedeniyle aile hekimi tarafından plastik cerrahi polikliniğine sevk edilen 29 yaşında bir kadındı. Hastanın özgeçmişinde demir eksikliği anemisi ve safra kesesinde polip haricinde bir özellik yoktu. Yapılan fizik muayenede çenesindeki lezyon 0,7 cm çapında kirli beyaz ve kahve renkli, burnundaki lezyon ise kirli beyaz renkli, kubbe şeklinde ve 0,3 cm çapındaydı. Lezyonların histopatolojik incemesi sonucu çenedeki lezyona intradermal nevüs, burundaki lezyona ise morfolojik özellikleri ve Faktör XIIIA zayıf (+) ve CD34 (+) boyanma paneli nedeniyle fibröz papül tanısı konuldu. Fibröz papülün benign veya malign deri lezyonlarıyla karışabilmesi nedeniyle olgunun literatür bilgileri eşliğinde tartışılması amaçlanmıştır.
Anahtar kelimeler: Fibröz papül, burun, genç kadın hasta

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Introduction

Fibrous papules are usually benign lesions seen in the nasal and perinasal areas of the face. (1,2,3,4,5,6,7,8,9,10). Their clinical appearance is not specific and they might be confused with benign-malign skin lesions such as nevus, basal cell carcinoma (BCC), wart, and soft-tissue or neural lesions, requiring more aggressive treatment, thus, especially their atypical forms might be misdiagnosed (1,4,5). It was reported

that the cases with high number of angiofibromas on the face might be related to tuberous sclerosis or type-I multiple endocrine neoplasias (8). Although the fibrous papule is classified in the same group with acral fibrokeratomes, penile papules, familial microvascular fibromas, and angiofibromas with adenoma sebaseum in recent years, all the fibrous papules are divided classified as classical, hypercellular, clear cell, pigmented, pleomorphic, epitheloid, inflammatory, and granular ones (1,2,4,7,9). It was aimed to present the this case because it can easily be confused with benign or malignant skin lesions.

Case Presentation

29-year-old woman who was referred to the plastic surgery polyclinic by her family doctor because of lesions on the chin and lateral side of the nose. In the patient's history, there was no characteristic other than iron deficiency anemia and polyp in gallbladder. In the physical examination, the off-white and brown-colored lesion on the chin was found to have 0.7 cm diameter and the lesion on the lateral side of the nose was found to have off-white color, dome shape, and 0.3 cm diameter. Then, both lesions were excised. The excised materials transferred to our department were macroscopically analyzed. The material excised from the chin had dimensions of 1.2x1x0.5 cm and there was an off-white/brown-colored dome-shaped lesion at the center. The material excised from the nasal region had the dimensions of 0.7x0.7x0.3 cm and there was an off-white lesion with 0.3 cm diameter at the center of material. In the histopathological analysis of both lesions, when immunohistochemical staining was applied, Melan A (+) staining was observed in tissue excised from the chin region, CD34 (+) staining in a focal region and vein wall, and SMA (+) in vein wall. Thus, the lesion was diagnosed with nevus. In the microscopic examination of the lesion excised from the nasal region, there were ganglion-like multinucleated giant cells, fibroblasts, and fibrovascular proliferation and dilatation on the surficial dermis (Figure 1-3). In immunohistochemical staining, CD68 was (+), Factor XIIIA was (+), SMA was (+), CD34 was (+), and S 100 was (-) (Figure 4-6). In the light of these findings, the case was diagnosed with fibrous papule.

Figure.1 *Ganglion-like giant cells, fibrovascular proliferation and dilatation, fibroblasts in superficial dermis, the flat epithelium that has polypoid appearance and multilayered at the surface (HE x 10).*

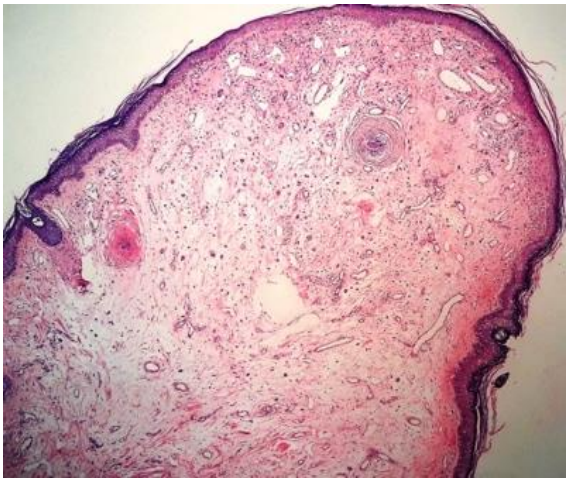


Figure.2 *Ganglion-like giant cells, fibrovascular proliferation and dilatation, fibroblasts in superficial dermis, the flat epithelium that has polypoid appearance and multilayered at the surface (HE x 20).*

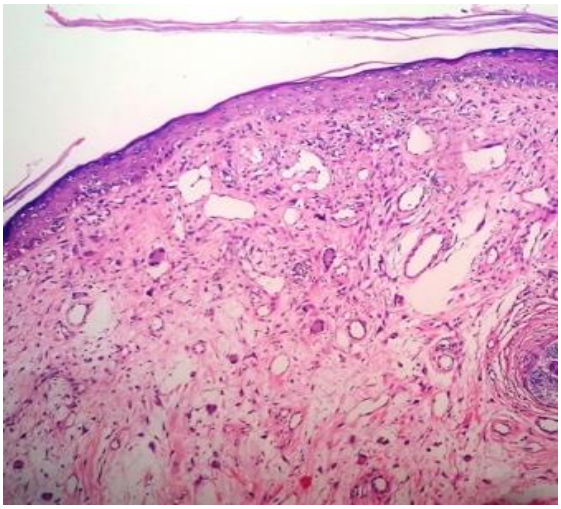


Figure.3 *A part of multilayered flat epithelium, spindle fibroblasts, blood vessels, and ganglion-like multinucleated giant cells on the surface (HE x 40).*

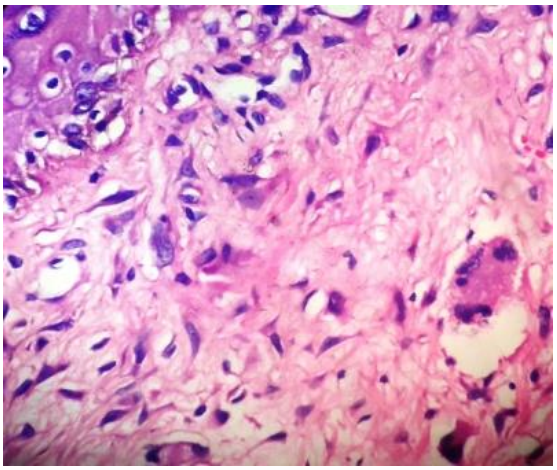


Figure.4 *CD34 positivity in spindle and multi nuclei (CD 34 x 40)*

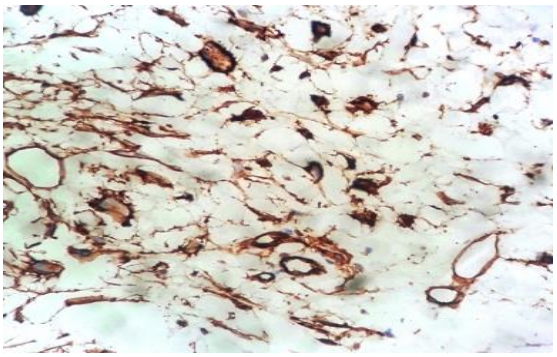


Figure.5 *Factor XIIIA positivity that is relatively strong in spindle cells and weak in multinucleated cells (Factor XIIIA x 40)*

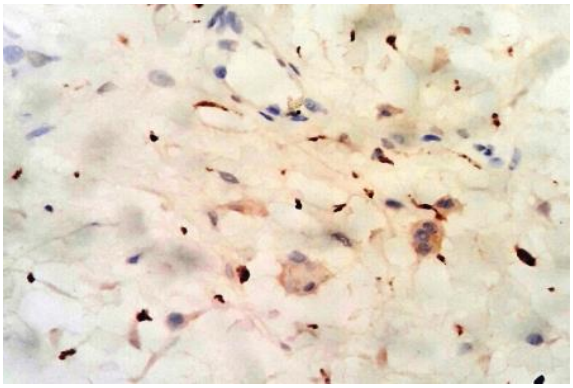
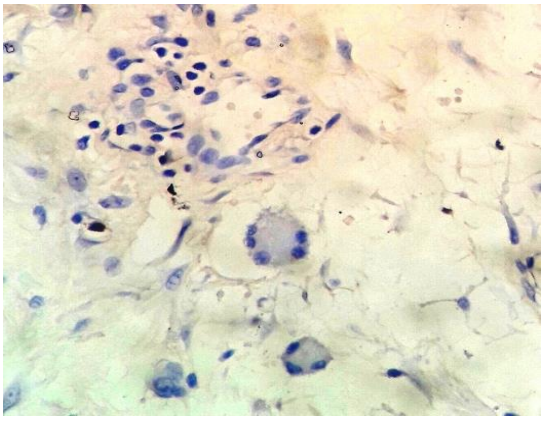


Figure.6 *S 100 negativity (S 100 x 40)*



Discussion

Fibrous papule was first defined as perifollicular fibroma by Zackheim and Pinkus in the year 1960 (2). The name "fibrous papule" became popular in the year 1965. The fibrous papules are skin-colored, asymptomatic, benign, and dome-shaped lesions generally seen on the nose and they affect both genders equally (1,2,3,4,5,6,7,8,9). From the clinical aspect, they might be confused with the benign and malign skin lesions such as BCC, adnexial tumors, pyogenic granuloma, nevocellular nevus, wart, acrochordons, intradermal melanocytic - desmoplastic nevi (DN), palisaded encapsulated neuroma, sclerosing perineurioma and sclerotic fibroma (1,2,4,5). Periungual angiofibroma can look similar to verruca vulgaris and subungual exostosis. Pearly penile papules can be confused with condyloma acuminatum and molluscum contagiosum (10). Clinically, sclerosing perineurioma usually presents as a solitary lesion; however, rare cases of multiple lesions have been reported, DN presents as a small and flesh-colored, erythematous or slightly pigmented papule or nodule that usually occurs on the arms and legs of young adults, palisaded encapsulated neuroma is a benign and solitary, firm, dome shaped, flesh-colored papule that occurs in middle-aged adults, predominately near mucocutaneous junctions of the face and sclerotic fibromas is well-demarcated, flesh-colored to erythematous, waxy papule or nodule with no site or sex predilection (2). The fibrous papules seen on the face are generally single but they might rarely be multiple and have diameters

smaller than 1 cm. The fibrous papules common seen on the nasal region are shiny, skin-color, hard, and dome-shaped papules with 1-5 mm diameter (8,9).

From the histopathological aspect, the fibrous papule consists of fibrous stroma with fibrous tissue-fibroblast proliferation, dilated blood vessels, and widespread, star-shaped, and multinucleated CD34 (+) cells (1,4,6,8,9). Moreover, the onion-like collagen structure is seen around the hair follicle. In some cases, the presence of melanocytes, number of which increases along the dermoepidermal junction and which are generally standing independently, and the inflammatory cells and lymphocytes distributed in dermis were reported (8,9). Different variants such as classical, hypercellular, clear cell, pigmented, pleomorphic, inflammatory, and granular types were identified (1,2,4,7,9).

The pathogenesis of fibrous papule is controversial and different etiologies were identified. It was claimed that they might be related with melanocytic nevus, hair follicle fibroma, proliferation of periadnexial dermal cells, and blood vessels (1). In fact, it has similarities with the spindle cell component of spitz nevus from the dermal cells and the increase of melanocytes throughout the basal membrane. Thus, they might be confused with melanocytic lesions (1). Besides that, in many studies, the theory of melanocytic origin was not supported because of the S-100 negativity in fibrous papule. Rather, it was claimed that these lesions originate from dermal dendrocytes stained with Factor XIIIA (1). From the immunohistochemical aspect, they might be confused with solitary fibrous tumor, DFSP, epithelioid fibrous histiocytoma, epitheloid sarcoma, neurofibroma, or meningioma (1). Actually, similar multinucleated giant cells might also be observed in epitheloid fibrous histiocytoma, giant cell angiofibroma, and orbital solitary fibrous tumor that may be seen on the face and it may cause histopathological diagnosis problems (1). However, the cases might be diagnosed by using the histopathological analysis, immunohistochemical findings, and clinical findings. In cases with many fibrous papules/angifibromas on the face, it is recommended to obtain a comprehensive clinical history, perform a comprehensive physical examination, and conduct necessary diagnostic tests in order to exclude the genodermatosis such as possible tuberous sclerosis or Type-I multiple endocrine neoplasms (8,9). Moreover, in literature, rare association of multiple fibrous papules with type-II neurofibromatosis, Cowden disease, Birt-Hogg-Dube syndrome, and Hornstein-Knickenberg syndrome was reported and it was claimed that it might be a variant of Birt-Hogg-Dube syndrome (8,9).

The stromal cells identified in fibrous papule might be star-shaped, spindle-shaped, bloated, or multinucleated (1). These cells might contain dark-colored and globular inclusion particles. In the present study, there were bloated, multinucleated, and ganglion-like cells. In these cells, CD68 was found to be (-), Factor XIIIA to be (+), SMA to be (-), CD34 to be (+), and S 100 to be (-). CD34-staining of giant multinucleated cells in fibrous papule has not been identified in previous cases. The staining profile in previous cases was found to be focal and the spindle cells on the bottom were apparent. The other lesions with uncommon floret-like multinucleated cells are include dermal pleomorphic lipoma and pleomorphic onychomatricoma and they are CD 34 (+). In previous ultrastructural studies, it was reported that there were plenty of similar giant endoplasmic reticulum similar in giant multinucleated cells in fibrous papule. Similar to the presence of Nissl particle in ganglion cells in routine histological analyses, there are plenty of them in granulated endoplasmic reticulum. Similarly, the diagnosis of pleomorphic adenoma is made upon the presence of extraordinary cells with apparent CD34 expression in dilated blood vessels (1). Although there are multinucleated and floret-like cells in typical pleomorphic adenomas, hyperchromatic or vesicular chromatin, multilobular nucleus, or another typical apparent atypia seen in pleomorphic fibroma are not observed. Moreover, there is no typical collagenous stroma (1). High number of ganglion-like giant cells might easily lead the histopathologist to different diagnoses such as ganglioneuroma or proliferated fasciitis, in which the ganglion cells are observed. Although the giant cells were rarely seen in similar studies before, their presence was reported in many studies identifying the fibrous papule but the presence of ganglion-like giant cells was not reported. In fact, the appearance of fibrous papule and that of other lesions, in which

multinucleated giant cells or ganglion-like cells are observed, are similar to each other. The common point of fibrous papules and other lesions with different appearances is the inclusions they contain (1).

In conclusion, fibrous papule is a melanocytic, and benign lesion, which generally courses much more aggressively, might be confused with soft tissue and neural lesions, require aggressive treatment, might accompany syndromes or genodermatosis when multiple, and should be distinguished for this reason.

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