

# Palmoplantar keratoderma and pleomorphic adenoma of submandibular salivary gland: A rare association in a pediatric patient

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## ABSTRACT

Palmoplantar keratoderma (PPK) is a rare disorder in which dry, thick patches of skin develop on the soles of the hands and feet. Our patient, an 11-year-old boy was undergoing treatment for progressive PPK by a dermatologist since the age of 1 year. He presented to us with swelling in the left submandibular region. Incision biopsy was suggestive of pleomorphic adenoma (PA). Patient was managed with complete surgical excision of the left submandibular gland. The association of both the diffuse and punctate forms of PPK with internal neoplasia has been described by a number of reports. We report the occurrence of PA of the submandibular salivary gland in a case of diffuse PPK. To the best of our knowledge this association has never been described before.

**Key words:** Palmoplantar keratoderma, pleomorphic adenoma, submandibular neoplasms

## INTRODUCTION

Palmoplantar keratoderma (PPK) (also known as Palmoplantar keratosis) is a rare disorder characterized by thickening of the skin on the palms and soles. PPK has been classified clinically according to the presence or absence of epidermolysis and the pattern of hyperkeratosis on the palms and soles. Diffuse, punctate, and focal forms of PPK have been described.<sup>[1,2]</sup>

80-90% of the salivary gland neoplasms arise from the parotid gland and only 4-8% arise from the submandibular gland.<sup>[3,4]</sup> Pleomorphic adenoma (PA) is the most common benign parotid neoplasm. PA of submandibular salivary gland is a rare occurrence and usually presenting in old age.<sup>[4]</sup>

The association of both the diffuse and punctate forms of PPK with internal neoplasia has been described in literature.<sup>[2,5-7]</sup> We report the occurrence of PA of submandibular salivary gland in a case of diffuse non-epidermolytic PPK. To the best of our knowledge this association has never been described before.

## CASE REPORT

An 11-year-old male was referred to us by a dermatologist for a gradually increasing swelling on the left side of neck. He was undergoing treatment for diffuse PPK by the dermatologist since the age of 1 year. Histopathology report of skin biopsy from the affected palm was suggestive of PPK. Family history of PPK was present. Incision biopsy was done for left submandibular swelling at another center. Histopathology report was suggestive of PA of submandibular salivary gland. On examination, there was swelling in the left submandibular region of size 4 × 3 cm, firm, non-tender, non-mobile with overlying scar of old biopsy incision [Figure 1]. Bimanual palpation was positive and there was no neuronal or functional disturbance of adjacent tissues. In addition the patient was having even, thick, symmetric, dry patches over the whole of the palm and sole. The patient was also having pseudoainhum

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of fingers. Teeth and oral mucosa was normal. All features were suggestive of diffuse non-epidermolytic PPK [Figure 2a and b].

Magnetic resonance imaging done after biopsy showed bulky left submandibular gland and a  $1.6 \times 1.6 \times 1.4$  cm well-defined lesion in the sublingual space [Figure 3].

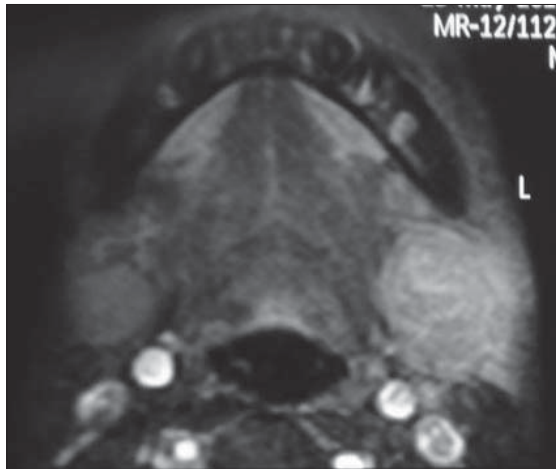
Complete excision of the left submandibular gland along with its duct was performed using submandibular incision incorporating the old scar. Histopathological examination was confirmatory of PA [Figure 4]. The patient did not have any neurological deficit or recurrence at 1 year of follow up. The patient had continued treatment for diffuse PPK with the dermatologist in the form of local application of 6% salicylic acid and oral retinoids.

## DISCUSSION

PPK is a rare skin disorder in which dry, thick patches of skin develop on the palms and soles. Classification



**Figure 1:** Left submandibular swelling with overlying scar of old biopsy incision



**Figure 3:** MRI showing well defined lesion in sublingual space

of PPK is confusing. The various forms of PPK can be divided into hereditary form with only skin problems, hereditary syndromes with PPK as an associated feature, and acquired forms. Based on distinct clinical patterns of epidermal involvement, hereditary form is further sub-classified into 3 types. (i) Diffuse type, in which there is uniform involvement of the palmoplantar surface. This pattern is usually evident within the first few months of life. (ii) Focal type, consisting of localized areas of hyperkeratosis mainly affecting pressure points and sites of recurrent friction. (iii) Punctate form, consisting of multiple small, hyperkeratosis papules, spicules, or nodules on the palms and soles.

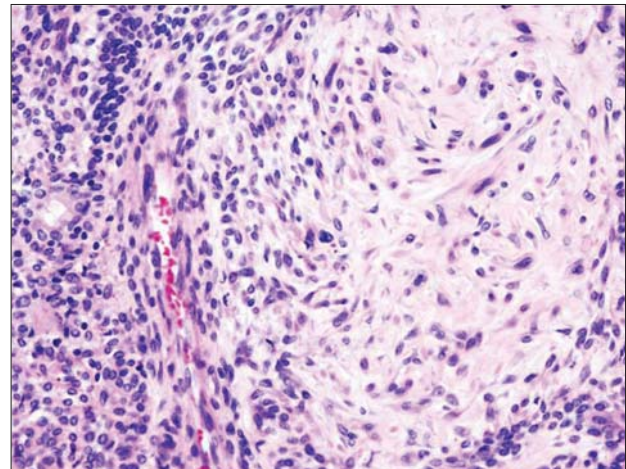
Diffuse hereditary PPK again can be classified as: (i) Diffuse non-epidermolytic PPK (ii) diffuse epidermolytic PPK (iii) progressive PPK.

Diffuse non-epidermolytic PPK is an autosomal dominantly inherited condition traced to KRT1 and KRT16 keratins. Onset of clinical features is usually within the first 2 years of life. Our patient presented for his dermatological problem in infancy. Clinical examination and histopathology was suggestive of diffuse non-epidermolytic PPK.

PA is also known as mixed tumor of salivary gland, which describes its pleomorphic appearance as opposed to its dual



**Figure 2:** (a) Hyperkeratosis of palms, (b) Hyperkeratosis of soles



**Figure 4:** H and E slide showing features suggestive pleomorphic adenoma

origin from epithelial and myoepithelial elements. PA is the commonest salivary gland tumor. PA most commonly occurs in the parotid gland and the submandibular gland is the second in order. Incidence is highest in the 3<sup>rd</sup>-6<sup>th</sup> decade of life, with female predominance.<sup>[8]</sup> PA usually presents as a painless, slow growing mass.

MRI is the gold standard for commenting on the origin and the extent of salivary gland tumors. FNAC or incision biopsy may be used initially for the diagnosis of PA.

Surgical excision is the treatment of choice in PA.<sup>[9]</sup> Twenty-five percent of untreated PAs undergo malignant transformation hence surgical excision is strongly recommended. Enucleation or superficial parotidectomy or total parotidectomy can be done in case of parotid gland PA. But in submandibular salivary gland complete excision is required.<sup>[9,10]</sup> Incomplete removal results in increased risk of recurrence. Complete excision of submandibular gland is challenging as it carries considerable risk of injury to the mandibular branch of facial nerve, hypoglossal nerve, and lingual nerve.

A possible association between PPK and internal malignancy has been noted by Herman in 1973. PPK is now considered to be associated with various malignancies including esophageal cancer, Hodgkin's disease, renal, breast, pancreatic, colonic adenocarcinomas, and malignant melanoma.<sup>[2,5-7]</sup> Non-epidermolytic PPK is associated with esophageal cancer. The disease locus (previously termed the "tylosis esophageal cancer gene" locus) has been mapped to 17q23-qter by linkage analysis. This region is located telomeric to the keratin 16 gene, in which mutations have been identified in focal PPK families.<sup>[2]</sup>

To date, no association between PPK and PA of submandibular gland has been observed. To the best of our knowledge, our patient is the only known case of diffuse

PPK with a rare pediatric neoplasm (PA of submandibular gland), to be reported in literature.

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