Digital pacinian neuroma in a patient with myeloma

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ABSTRACT

We present a case of Pacinian Neuroma (PN) associated with multiple myeloma (MM) in a 57-year-old man with a 6-month history of painful swelling of his fingers. Physical examination revealed definite palpable nodules and moderate swelling over the first phalanx of all his fingers. A biopsy confirmed the diagnosis of PN. The patient reported chronic back pain. Radiologically, a severe demineralization was found. MM was suspected. The results of complete blood count and biochemistry panels were within normal limits. Otherwise, immunoelectrophoresis demonstrated monoclonal light chains in blood and urine. Bone marrow biopsy was consistent with MM. The patient successfully underwent an autologous stem cell transplant with good response. A slight decrease in the size in PN was noted. To the best of our knowledge, no case of PN associated with MM had been reported. This occurrence may be accidental. However, further studies are required in order to explain this association.

Key words: Pacinian corpuscles; Pacinian neuroma; Multiple myeloma

INTRODUCTION

Pacinian corpuscles (PC), a rapidly adapting pressure-sensitive mechanoreceptor, found in several organs and are mainly distributed in the dermis of the fingers and palm of the hand. Reactive enlargement of these PC is known as Pacinian neuroma (PN): an extremely painful condition with only few cases reported in the literature. It is characterized by formation of painful nodules in connective tissue of the hand. The etiopathogenesis is still not clear. However, it is of non-malign nature and frequently reported after local or repetitive trauma. We present here a case in which PN was associated with multiple myeloma.

CASE REPORT

A 57-year-old previously healthy man presented with a 6-month history of severe pain and swelling located over the volar surface of his fingers. He reported no previous history of trauma or hand injury. He didn’t present any neurological symptoms. Physical examination revealed definitely palpable nodules and moderate swelling over the first phalanx of his left thumb and all his fingers (Fig. 1).

A biopsy specimen was performed. It revealed multiple hyperplastic Pacinian corpuscles in the lower dermis and subcutaneous fat (Fig. 2). A diagnosis of PN was made. The patient also reported chronic back pain and muscle weakness. Lumbar spine X-ray showed a severe demineralization. MM was suspected. On laboratory evaluation, the results of complete blood count and biochemistry panels were within normal limits. There was no anemia, hypercalcemia, renal impairment or lytic bone lesions. Otherwise, immunoelectrophoresis demonstrated monoclonal light chains in blood and urine. Bone marrow biopsy was obtained and was consistent with plasma cell myeloma. The patient successfully underwent an autologous stem cell transplant with a very good response. Two months after his myeloma remission,
pain was slightly alleviated with a slight decrease in the size in PN.

DISCUSSION

PN is an extremely rare feature. To date, only 75 cases have been reported. It is defined as hyperplasia or hypertrophy of PC corresponding to tactile receptors located in the deep dermal and subcutaneous of palmar and plantar skin. The PC is a single myelinated nerve fibre, except for the terminal region within the capsule which has a characteristic onion-like cut surface. Almost all reported cases of PN were clinically characterized by painful lesions related to digital nerves of the hands in middle-aged adults.

A case of PN presenting as congenital macrodactyly of the digit had been reported [1]. Extra digital PN is exceptional. It can be localized to the visceral organ or mucosa [2]. Histologically, PN shows mature Pacinian corpuscles of increased size or number, in association with degenerative changes and fibrosis of the adjacent nerve.

The exact pathogenesis of PN remains unknown. Local trauma or surgery has been proposed as a predisposing factor in about 50% of cases [3]. It has been hypothesized that this local injury may disturb the relationship between the corpuscle and arteriovenous anastomoses resulting in PN [4,5]. Some authors propose that PN of the hand may be considered as an occupational injury [6]. In other cases of PN without trauma, growth of pre-existing Pacinian corpuscles can be due to unknown causes [7]. PN has been reported associated with glomus tumors or Dupuytren’s disease [8]. Rare cases occurred in patients with neurofibromatosis type 1 [9].

The discovery of MM was concomitant with the appearance of PN. Myeloma is most often accompanied by cutaneous lesions such as cutaneous plasmacytoma. Other dermatoses such as leukocytoclastic vasculitis, urticaria, autoimmune bullous diseases, and pyoderma gangrenosum have been reported [10]. The PN has never been described in patients with MM. Although there is a pain regression of lesions after the treatment of myeloma, a fortuitous association cannot be eliminated.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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