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Palmar fasciitis and polyarthritis syndrome—Systematic literature review of 100 cases

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ABSTRACT

Objective: To analyse clinical, laboratory, and imaging characteristics of all patients with palmar fasciitis and polyarthritis syndrome (PFPAS) described in the literature.
Method: Comparison of the clinical presentation of one patient with acute onset of PFAPS with 99 other published cases identified through a PubMed literature research.
Results: Since the original description in 1982 by Medsger et al., there have been numerous case reports and small case series in the literature. In total, 73 articles in English, French and Spanish language were included in the analysis. PFPAS is a rare but characteristic paraneoplastic syndrome in rheumatology. Its distinct clinical feature is a painful swelling of both the hands caused by an inflammation of the palmar fascia, tendon sheaths and small joints of fingers and wrist, and flexion contractures develop rapidly. Since the subcutaneous tissues become indurated and hard, the illustrative term "woody hands" was coined. The most frequent underlying malignancy is ovarian cancer but adenocarcinomas of the breast, gastrointestinal tract and other organs can also cause this syndrome. A helpful diagnostic procedure in order to identify the nature of the underlying malignancy in many cases has been the determination of various serum tumour markers. In cases when a complete removal of the malignancy is possible, PFPAS can also undergo complete remission.
Conclusions: Knowledge of the distinct features of this rare paraneoplastic syndrome facilitates early diagnosis and potentially life-saving therapeutic interventions.

Paraneoplastic syndromes are cancer-associated diseases or symptoms that occur distant from the underlying malignancy. They are not directly caused by the tumour or its metastases but are mediated by humoral factors, such as hormones and cytokines, or they are a consequence of the immune response against tumour cells. Various musculoskeletal conditions can be triggered by a malignant process: periostitis, arthritis, fasciitis, myositis, panniculitis or vasculitis can be a paraneoplastic symptom and therefore of interest to the rheumatologist [1]. The observation of a patient with acute severe palmar fasciitis and polyarthritis prompted us to conduct a systematic literature research to analyse all patients from case series and single case reports described up to this date.

The first description of a patient with palmar fasciitis and arthritis of finger joints in association with metastatic ovarian carcinoma by Bremer [2] dates back to 1966. However, despite its symmetric presentation, these symptoms were then designated "shoulder–hand syndrome", a variant of reflex sympathetic dystrophy or complex regional pain syndrome. It was not until 1982, when Medsger et al. [3] reported about six postmenopausal women, who developed very similar symptoms as a consequence of malignant ovarian tumours. In this article the term "palmar fasciitis and polyarthritis syndrome" (PFPAS) was created, which allowed to recognise this as a rare but characteristic paraneoplastic disease entity in rheumatology.

Materials and methods

To analyse all published patients with PFPAS, we conducted a systematic literature research. A PubMed search using "palmar" and "fasciitis" for the years 1982–2013 retrieved 76 articles. All were screened in full-text version and 58 were included in the analysis, because they reported individual patient cases. All relevant articles were retrieved and additional references quoted in these articles were checked. This led to the inclusion of 14 additional full-text articles in English, French and Spanish. These 72 articles together with the initial report by Bremer [2] describe the clinical characteristics of 99 patients with PFPAS. All cases were reviewed as full-length articles in original language, and both the

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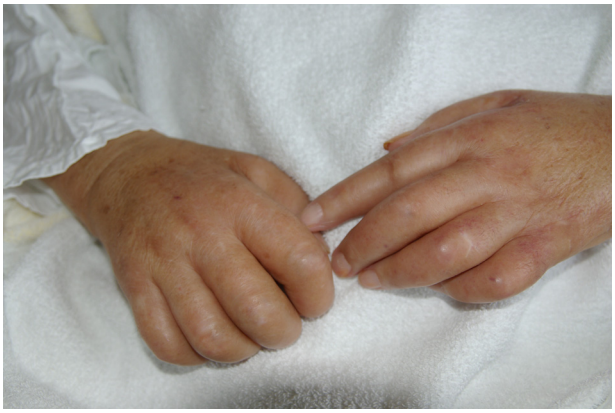

Fig. 1. Dorsal aspects of hands of a 73-year-old patient with a sudden onset of extremely painful bilateral hand swelling with rapid development of flexion contractures.

authors classified these cases as PFPAS based on the major clinical symptom of palmar fasciitis supported by additional clinical, laboratory, imaging and histopathological data. This makes the patient recently seen in our department the 100th observation of this syndrome in the literature. In 87 patients, PFPAS was associated with an underlying malignancy [2–67], in 10 it was attributed to benign conditions [68–70] or to medication [71] and in three it remained idiopathic [72–74]. The following results will analyse the findings in all 87 cases of paraneoplastic PFPAS, the 13 cases without malignant disease will be discussed separately below.

Results

Case report

A 73-year-old female farmer experienced an acute onset of very painful symmetrical swelling of both the hands with stiffness and diffuse arthralgias in the elbows and the knees. In addition, she suffered from fatigue and reported a weight loss of 2 kg within the last 6 weeks. A presumptive diagnosis of initial systemic sclerosis was made but therapy with 50-mg prednisone per day had no effect at all and flexion contractures with bilateral palmar induration developed rapidly (Figs. 1 and 2). Magnetic resonance imaging showed palmar fascial and peritendinous signal enhancement and gadolinium uptake in tendon sheaths of flexor and extensor tendons.

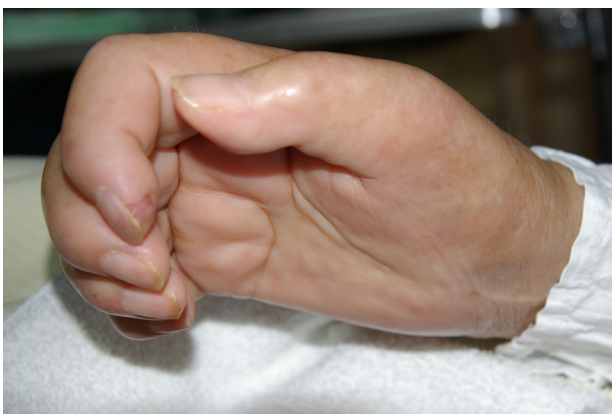

Fig. 2. The palmar nodular fasciitis resembles Dupuytren's contracture but is in contrast much more severe and inflammatory. The atrophic scar at the tip of the index finger stems from an earlier injury and is not caused by the PFPAS disease process.

Two months after the initial onset of musculoskeletal symptoms, CT examination showed a mass of the left ovary with signs of peritoneal and omental metastases. Hysterectomy and bilateral ovarectomy with removal of peritoneal, omental and para-aortal metastases revealed a poorly differentiated serous adenocarcinoma. The patient received postoperative chemotherapy with carboplatin and paclitaxel, but this did not influence her musculoskeletal symptoms; the tumour progressed and she died only about 2 months later.

Literature review

Musculoskeletal symptoms

In most of the reported cases, PFPAS presents with a sudden onset of diffuse painful swelling of both the hands with marked stiffness. Later nodular thickening of the palmar fascia develops similar to Dupuytren's contracture, but much more severe. Overall, 20% of the reported cases exhibit a similar involvement of the plantar fascia. The skin of the involved area is shiny and tight, and some case reports describe an erythematous or acrocyanotic discoloration. However, there is only one description of Raynaud's phenomenon [43], sclerodactyly is absent and capillary microscopy findings are normal. Some authors describe the palpatory findings in an advanced stage with the illustrative term “woody hands” [29,42,56]. Occasionally a “groove sign” of involved areas of the hands can be seen, which is an indentation of the skin over superficial veins, when the extremity is elevated [35,52]. The bilateral “woody hands”, flexion contractures and nodular palmar fasciitis of our patient are shown in Figures 1 and 2.

The polyarthritis presents with arthralgias and synovitis of metocarpophalangeal and proximal interphalangeal joints and wrists. Progressive flexion contractures lead to a rapid loss of hand function. PFAPS in most cases shows symmetrical involvement of fascia and joints, but occasionally strictly [33,59] or predominantly [46,55,65] unilateral manifestations have been reported. Arthritides of other joints are frequent but usually considerably milder than those of hands and fingers. The shoulders are most frequently involved, which can present as adhesive capsulitis with a markedly reduced range of motion. Before the concept of PFPAS as a separate disease entity was generally accepted, often these patients were diagnosed with “shoulder–hand syndrome” [2,6,9] or reflex sympathetic dystrophy [7,10,13,17]. In eight patients carpal tunnel syndrome was present as a complication of their wrist involvement [3,23,33,35,40,41,43]. Other frequently involved joints are the elbows, knees and ankles. In four patients with paraneoplastic PFAPS, arthrocentesis was performed because of knee synovitis with effusion [33,35,38,58].

Type of underlying malignancy and epidemiology

In 36.8% of women, by far the most frequent tumour type in paraneoplastic PFPAS is ovarian adenocarcinoma. Ovarian and breast cancer together with malignancies of other female reproductive organs are the underlying cause in more than half of all published cases (Table 1). This accounts for the fact that PFPAS has been described in females more than four times more frequent than in males. This observation has often been used as an argument for a pathophysiological role of female sex hormones in the pathogenesis of this paraneoplastic syndrome. However, when all patients with malignancies of female reproductive organs, breast cancer and prostate cancer are excluded from the analysis there remains only a small female preponderance of 1.3–1 for all other tumour types. Interestingly and in contrast to some other paraneoplastic syndromes, only five PFPAS patients with hematolymphatic malignancies have been described.

In 73.8% of all cases, by far the most frequently occurring histological type was adenocarcinomas, for which detailed