Vertebral lesions associated with palmoplantar pustulosis

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Summary. *Spinal lesions were reviewed in 9 patients (3 men and 6 women) who presented with palmoplantar pustulosis. Cervical lesions were found in 3 patients and lumbosacral involvement in 7. The duration of disease averaged 4.8 years. Seven patients had associated sterno-costo-clavicular hyperostotic lesions, and all showed abnormalities in blood tests which suggested chronic inflammation. Vertebral lesions were assessed as a spondylodiscitis type in 4, single level syndesmophyte-forming type in 1, multiple hyperostosis type in 4, and an ivory vertebra type in 1. Five patients were treated conservatively, and 4 with progressive spondylodiscitis type lesions underwent curettage and anterior interbody fusions. At follow-up, all patients improved in both spinal and cutaneous presentations, and 4 who underwent surgery and 1 with tonsillectomy attained complete cure. The vertebral manifestations of palmoplantar pustulosis may vary, but rapidly progressive lesions need surgery for favourable early improvement from the vertebro-cutaneous symptom complex.*

Résumé. *Revue de 9 patients présentant des lésions vertébrales associées à une pustulose palmo-plantaire. Il s'agissait de 3 hommes et 6 femmes avec une maladie évoluant en moyenne depuis 4,8 ans. Des lésions cervicales ont été trouvées 3 fois, et des lésions lombo-sacrées 7 fois. 7 patients avaient des lésions hyperostotiques de la ceinture scapulaire, ce qui est classique, et tous présentaient un syndrome biologique inflammatoire.*

Les lésions vertébrales étaient une spondylodiscite 4 fois, une syndesmophytose d'un niveau, 1 fois, une hyperostose étendue 4 fois et une vertèbre ivoire 1 fois. 5 patients ont eu un traitement conservateur et 4 qui présentaient une spondylodiscite ont été opérés par curettage et arthrodèse vertébrale antérieure. Au dernier recul, tous les patients ont eu une amélioration tant de la symptomatologie vertébrale que des lésions cutanées et 5 peuvent être considérés comme guéris: les 4 opérés pour spondylodiscite et 1 opéré par amygdaléctomie. Les lésions vertébrales associées à la pustulose palmo-plantaire sont variables mais si elles sont d'évolution rapide, le traitement chirurgical est utile pour l'amélioration du syndrome cutanéovertébral.

Introduction

Palmoplantar pustulosis (PPP) is seen infrequently in association with nonspecific chronic multifocal osteomyelitis [1, 2, 3, 26]. Most reports have suggested that sterno-costo-clavicular hyperostosis or nonspecific osteitis is a significant aetiology of PPP or, at least, is an important factor contributing to the cutaneous manifestations [4, 5, 13, 15, 16, 23, 31]. This osseo-arthro-cutaneous syndrome shows unique clinical manifestations which may correlate with each other during its lengthy course [8, 9, 14, 18]. Many reports have concentrated on the upper portion of the anterior thorax, but the vertebral involvement in PPP can be of clinical significance [17]. We present the clinical findings in vertebral lesions associated with PPP.

Table 1. Clinical presentation

Patient No.	Age/sex	Duration of disease (years)	Other associated lesions	Blood examination at initial visit				Follow-up (years)	Condition ^e at follow-up
				ESR ^a	CRP ^b	WBC ^c	ALP ^d		
1	47/M	2.5	Tonsillitis, bil. sterno-clavicular osteitis	24/ 45	2+	4,800	176	9.9	A
2	56/F	1.3	Tonsillitis, teeth caries, bil.sterno-costo- clavicular osteitis	48/ 61	3+	8,200	155	8.7	A
3	53/M	8.0	Tonsillitis, IgA-type nephropathy	50/ 74	3+	8,900	146	7.3	A
4	61/F	3.5	Tonsillitis, bil.sterno-costal osteitis	38/ 56	2+	7,500	215	4.7	B
5	65/F	4.8	Bil. sterno-clavicular osteitis, chronic otitis media	28/64	2+	5,600	168	3.9	C
6	56/F	10.0	Bil. sterno-costal osteitis	119/140	5+	10,200	268	2.8	A
7	46/F	3.8	Tonsillitis, bil. sterno-clavicular osteitis	55/ 80	3+	8,300	240	1.7	A
8	44/F	0.3	Rt. sterno-clavicular osteitis	48/ 76	3+	9,700	186	1.4	C
9	53/M	9.0	Lt. chronic femoral osteomyelitis (non-specific)	46/ 75	3+	8,400	215	0.9	C

^a Erythrocyte sedimentation rate (mm, per 1 h/2 h).

^b C-reactive proteins (g×100 ml⁻¹); 1+, 0.77~1.66, 2+, 1.67~3.35, 3+, 3.36~6.41, 4+, 6.42~9.68, 5+, 9.69~13.83, 6+, 13.84 or more.

^c White blood cell count (mm⁻³);

^d Alkaline phosphatase (normal value, 80–280 IU×ml⁻¹).

^e Grade A, spinal symptoms disappeared without recurrence of skin rash; grade B, spinal symptoms markedly improved and no recurrence of cutaneous rash; grade C, spinal symptoms decreased but remain, and skin rash occasionally recurs; grade D, no improvement in both spinal and cutaneous presentation; grade E, both spinal and cutaneous symptoms and signs progressed

Patients and methods

We have reviewed 9 patients (3 men and 6 women) who presented with vertebral and cutaneous manifestations of PPP. Their average age was 47 years, and the average duration of PPP was 4.8 years. They had been treated medically for the presumed diagnosis of pyogenic spondylitis. Some patients with a long history of PPP had followed a recurrent course, with aggravations and remissions of their cutaneous and spinal problems.

The vertebral lesions showed a wide spectrum of radiological features. Even in the same patient, radiographs and magnetic resonance imaging (MRI) showed multimodal appearances resembling such clinical entities as acute or chronic pyogenic spondylodiscitis, ankylosing spondylitis, and diffuse idiopathic skeletal hyperostosis (DISH). We classified the vertebral lesions into the following subtypes:

Type I, a lesion similar to pyogenic spondylodiscitis, with osteolytic lesions followed by an osteosclerotic reaction.

Type II, unilateral or bilateral marginal syndesmophyte formation with some intracorporeal hyperostosis, or an arched bony bridging over several vertebral corners.

Type III, single- or multi-level paravertebral syndesmophyte formation or paravertebral ossification resembling DISH.

Type IV, diffusely spanned intracorporeal hyperostosis.

The vertebral lesions were also categorized as acute and progressive, subacute, or chronic. Five patients, 2 with cervical and 3 with lumbosacral involvement, subsequently underwent anterior curettage of the vertebral lesions followed by autogenous iliac grafting because of resistance to conservative management and/or progression of vertebral lesions. One

patient required transpedicular biopsy to exclude a metastasis. The resected and biopsied specimens were submitted for histological and bacteriological examination. Some patients showed bone and joint involvement other than in the spine, but these lesions were clinically and radiologically less significant than those in the vertebrae.

At follow-up (average 4.2 years), the vertebro-cutaneous symptom complex was rated into five grades:

Grade A, both spinal and cutaneous symptoms cured.

Grade B, spinal symptoms markedly improved with no recurrence of skin rash.

Grade C, spinal symptoms decreased but persistent, and skin rash occasionally recurring.

Grade D, no improvement in both spinal and cutaneous manifestations.

Grade E, both spinal and cutaneous symptoms and signs worse.

Results

Clinical presentation and blood examination

Table 1 lists the clinical data. Seven patients had sterno-costo-clavicular lesions and 5 had tonsillitis. Patients 1 and 6 had a radiculopathy. All showed an elevated erythrocyte sedimentation rate and increased C-reactive protein levels. Bacteriological studies confirmed that the palmoplantar pustules were aseptic; all patients had a negative

Table 2. Distribution of spinal lesions and their management

Patient No.	Symptoms and signs	Level of involvement	Type of lesion ^a and progress	Management
1	Neck pain, bil. radiiculopathy	C5–6	I, acute and progressive	Curettage and anterior interbody fusion, NSAIDs ^b
2	Neck pain	C5–6	I, subacute and progressive	Curettage and anterior interbody fusion, NSAIDs
3	Lower back pain	L1–2	I, acute and progressive	Curettage and anterior interbody fusion, NSAIDs, antibiotics (i. v.) ^c
4	Lower back pain	L3, L5	II, chronic	NSAIDs, antibiotics (p. o.) ^d
5	Lower back pain	L2, L5	IV, chronic	NSAIDs, antibiotics (p. o.)
6	Lower back pain, left sciatica	L5–S1	I, acute and progressive	Curettage and anterior interbody fusion, NSAIDs, antibiotics (i. v. and p. o.)
7	Neck stiffness, lower back pain	C4–C7, L4–5	III (C4–C7), chronic; III (L4–5), chronic	Tonsillectomy, NSAIDs, antibiotics (p. o.)
8	Middle back pain	L2–3	III, chronic	NSAIDs
9	Lower back pain	L5–S1	III, chronic	NSAIDs, antibiotics (i. v. and p. o.)

^a Type I, spondylodiscitis type; type II, unilateral or bilateral syndesmophyte formation and slight intracorporeal hyperostosis; type III, multiple diffuse idiopathic skeletal hyperostosis type; type IV, lesion resembling ivory vertebra or hyperostotic metastasis.

^b Non-steroidal anti-inflammatory drugs.

^c and ^d Intravenous and per os administration

rheumatoid factor and lupus erythematosus tests and were negative for the human lymphocyte antigen B27. Two patients showed a mildly elevated antistreptolysin-O level.

Radiologic findings

Three patients had cervical lesions and 7 had lumbar spine involvement (Table 2). Radiological findings varied greatly from Type I (Fig. 1) to Type IV (Fig. 2). One patient had a typical and asymmetrical syndesmophyte lesion with intracorporeal hyperostosis (Fig. 3). The vertebral lesions were assessed as Type I in 4 patients, Type II in 1, Type III in 4, and Type IV in 1 case. The Type I lesions in patients 3 and 6 showed low intensity signals on T1-weighted images (Fig. 4), and high intensity signals on a T2-weighted sequence. The Type IV lesion demonstrated a combination of iso- and hypo-intensity signals on the T1-weighted MRI sequence (Fig. 5), and iso- to high intensity areas in T2-weighted images.

Histological examination

The biopsy from patient 5 and surgically resected specimens were examined histologically. The Type I lesion (patients 1, 2, 3 and 6) showed non-specific granulation tissue with significant infiltration of mostly lymphocytic small round cells,

consistent with osteomyelitis. Samples from the Type IV lesion showed a few small round cell infiltrations, derangement of the bony trabeculae and moderately increased osteoid activity.

Assessment at follow-up

Five patients were managed conservatively. At follow-up, 1 was categorised as Grade A, 1 as Grade B, and the other 3 as Grade C. The patient who underwent tonsillectomy remained in Grade A. All 4 patients who underwent surgery for the vertebral lesions were Grade A with solid fusions.

Discussion

An unknown immunological reaction relating to longstanding focal infection or some inflammatory process appears to contribute to the cutaneous symptoms of PPP [10, 17, 23, 25]. Among various sites for infection foci in bones and joints, a hyperostotic lesion in the sterno-costo-clavicular region has been implicated most frequently [4, 7]. A small number of these cases occur in association with vertebral lesions [11, 19, 30]. This osseo-cutaneous symptom complex may undergo intermittent remissions and aggravations during its lengthy clinical course, occasionally showing loss of bone and joint integrity as well as neurological

complications. The clinical and radiological features must be determined to permit appropriate patient care [12, 20].

The incidence of bone and joint involvement in PPP is more than 80% in the anterior chest region, 20–37% in the spine, and less than 22% in the sacroiliac joint and the pelvis [21]. A type of long bone osteomyelitis is assumed to be a cause in less than 3% [21, 31]. Vertebral lesions range from images consistent with acute spondylodiscitis to the multi-level ankylosing spondylitis of DISH and are non-specific to this osteo-cutaneous symptom complex. Sonozaiki et al. have noted small areas of sclerotic change in the anterior and anterolateral areas of the vertebrae, which are different to those seen in the acute phase of spondylodiscitis [32].

Fig. 1. Plain cervical radiograph of Patient 1, showing osteolytic changes at C5–6, similar to those in the acute phase of spondylodiscitis

Fig. 2. Plain lumbar spine radiograph of Patient 5, demonstrating diffusely spanned hyperostotic changes within the vertebral body at L2 and L3

Fig. 3. Radiograph showing the Type II lesion (L3 and L5) seen in Patient 4

Fig. 4. Magnetic resonance imaging showing low intensity signals on T1-weighted image (TR, 300 ms and TE, 20 ms), at the level of L5 and S1 (Patient 6)

Fig. 5. Magnetic resonance imaging (Patient 5) showing irregularity of signal changes on T1-weighted image (TR, 380 ms and TE, 20 ms) at the level of L2 and L5

Kawai et al. have reported some patients with vertebral lesions similar to those seen in psoriatic arthropathy and Reiter's disease [21]. Resnick and Resnick et al. distinguished the lesions from those associated with DISH [27, 28, 29]. DISH is often associated with ossification of the posterior longitudinal ligament and ligamentum flavum, causing neurological compromise, without evidence of inflammatory changes in the spine. Ankylosing spondylitis frequently shows chronic hyperostotic changes and multiple bony bridges over the anterior vertebral columns, resembling the Type II vertebral lesion in our classification, but can be differentiated from PPP serologically. All 9 patients were HLA-B27 negative. At present, we consider that the vertebral lesions represent variable forms of chronic osteomyelitis, relating to aseptic cutaneous manifestations.

The management of this symptom complex requires a thorough clinical and haematological assessment including dermatological evaluation [6, 9, 22, 24]. Slowly progressing vertebral lesions, or a seemingly chronic phase lesion such as in Types II, III and IV, should be medically treated. When the vertebral lesion is acute and rapidly progressive, surgical intervention is recommended to diminish the cutaneous presentation and regain spinal stability. Four of nine patients in our series required surgery, resulting in early satisfactory relief of symptoms. In one patient (patient 7), vertebral lesions at both the cervical and lumbar levels were assessed as inactive or latent, and consequently she improved following surgical drainage of acute pyogenic tonsillitis. The vertebral lesion seen in PPP often shows a prolonged course, and surgery is indicated only if the lesion shows progressive osteomyelitic changes associated with significant cutaneous and spinal symptoms.

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