Facial Paralysis during Sarcoidosis: About Seven Cases

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Abstract

The aim of this work was to specify the frequency and the peculiarities of facial paralysis (FP) during in Tunisian patients. It’s a retrospective and descriptive study during a period of 14 years concerning patients affected by sarcoidosis and having a facial nerve involvement. Among 160 patients affected by systemic sarcoidosis, we counted seven cases of FP (3.1%) mainly women with a mean age at the diagnosis of facial nerve involvement of 52.2 years. FP revealed the disease in six cases. It was fluctuating in three cases and left in four cases. The diagnosis of Heerfordt syndrome was retained in one case. It was essentially associated to mediastinal ganglionic involvement or to lung interstitial pneumopathy. Oral corticosteroid therapy allowed fast regression of FP in a mean delay of 3 months. The facial nerve involvement during sarcoidosis is rare. It is more frequent in women and it would be most of the time revealing and of left seat. The response to corticosteroids is fast.

Key words

Facial palsy, Sarcoidosis, Heerfordt’s syndrome

Introduction

Neurological damage in sarcoidosis is quite rare, observed in 5 to 10% of cases. It is polymorphic and can affect the peripheral nervous system, the central nervous system and the meninges. The involvement of one or more cranial nerves is noted in 24 to 73% of neuro-sarcoidosis, especially in black race and especially for facial nerve (20-50% of cases), causing peripheral, rocking, and willingly recurrent FP [1-3]. The objective of our work was to clarify the frequency and peculiarities of facial nerve damage during sarcoidosis in Tunisian patients.

Patients and Methods

This is a retrospective descriptive study from 2000 to 2019 of patients with sarcoidosis with facial nerve damage. The diagnosis of sarcoidosis was retained in front of clinical, biological, radiological arguments, with or without histological evidence. Other causes of periphera IF Pwereruled out.

Results

Of the 160 patients with systemic sarcoidosis, seven were diagnosed with FP (3.1%). There were 5 women and 2 men. The mean age at diagnosis of sarcoidosis was 47.3 years and at diagnosis of FP was 52.2 years. FP was indicative of the disease in 6 cases. It occurred 3 years after the diagnosis of the disease in case. It was rocking and recurrent in three cases respectively and left seat in four cases. The beginning was brutal in all cases. FP was associated with: Mediastinally lymphnode involvement in four cases, interstitial pneumopathy in four cases, granulomatous anterioruveit is in three cases, panuveitis in two cases, central neurological involvement in 3 two cases, meningeal involvement in a case, parotiditis in 1 case and erythem anaosom in 1 case.

The diagnosis of Heerfordt syndrome was retained in one case. In biology, there was hypercalciuria and hypercalcemia in 3 cases respectively. Lymphopenia was noted in 4 cases. The conversion enzyme assay in 7 patients was high in 4 cases. The salivary gland biopsy showed an epithelial granuloma with out caseousnecros is in 3 cases. Cerebral MRI showed a hypersignal of the white substance supra-tentorial non-specific in one case and a brain damage leptomeninia under tentoriel and contrast taking shooting of cerebellar and frontal ponto angles in another case. Oral corticosteroids allowed rapi d regression of FP in six cases and this within an
average of 3 months. One patient had a recurrence of the contralateral disease despite treatment. The clinical, paraclinical and evolutionary characteristics of the seven patients were reported in (Table 1).

**Discussion**

Our study describes the clinical, paraclinical and evolutionary features of facial nerve damage during sarcoidosis in a group of Tunisian patients. In this retrospective study, we identified seven FP cases from a total series of 160 patients (3,1%). In the Tunisian series, neurological damage during sarcoidosis was noted in 9.6 to 14% of cases [4,5]. These frequencies are very close to other foreign series: Neurological complications are noted in 5-15% of patients [6-9]. In the Tunisian series, paralysis of the cranial nerves was reported in 5% of cases with involvement of the 2nd, 5th, 7th and 8th cranial pairs [4]. However, the frequency of facial nerve damage was not estimated. In the other series, NS frequently manifests itself in cranial nerve damage in 24-73% of cases (mainly facial nerve and optic nerve) [1-3,10]. Systemic sarcoidosis affects both sexes with a female predominance. In our series, there was a clear female predominance. The meanage of sarcoidosis patients is generally between 20 and 40 years with a second peak in frequency between 45 and 65 years [11]. The meanage of our patients is at the second peak. Damage to the facial nerve, resulting in peripheral FP, is often indicative of the disease [3,12]. This result was confirmed in ourseries. Indeed, the FP had revealed sarcoidosis in sixcases. In one case, it occurred during evolution.

On the other hand, the FP during sarcoidosis would willingly flip over and repeat [3,12]. In our series, the FP was relapsed and rocking in only a third of the cases. This could be explained by early diagnosis and treatment. Other FP-associated systemic disorders were dominated by interstitial pulmonary involvement, mediastinally oropharyngeal involvement, and ocular involvement. The facial nerve involvement can fallinto heerfordt syndrome combining peripheral facial paralysis, bilateral anterioruveitis it is, bilateral parotiditis and fever. During this syndrome, the FP is very common it is almost of a latera and complete; it is sometimes bilateral. FP is often attributed to concomitant parotiditis compression.

However, it does not always evolve in parallel with this one [13]. The FP can sometimes precede eparotiditis is, as is the case withour patient. In addition, it is often unilateral while parotiditis is bilateral. F.P. can occur in the absence of parotiditis [10]. The lesion process would likely beneuritis of the face. Heerfordt syndrome has some similarities with sarcoidosis, of which it is only a particular form; Reticulo-endothelios is of viral origin is involved while others have considered a typical tuberculosis [13]. In sarcoidosis, FP may also be linked to VII impairment with in a granulomatous location of the temporal bone but in this case, association with auditory signs is common and will lead the diagnosis of FP during sarcoidosis require seliminatingo the retiologies. Idiopathic or refrigerated facial paralysis is the most common cause but this fact should not exempt to look for other causes, (Traumatic, toxic, infectious, tumor, inflammatory, congenital) The etiological approach is based primarily on a rigorous examination and clinical ENT and neurological examination that will guide the complementary biological, radiological and cochleo-vestibular investigations and make the diagnosis. Before the association of a parotiditis, a fever and an iridocyclitis (Heerfordt syndrome), neurological signs and/or pulmonary or chronically phadenopathy, other examinations will be performedmainly.

**Table 1:** Clinical, para-clinical and evolutionary characteristics of the facial paralysis.

| Case | Age at the diagnosis of PF | Sex | Characteristics of PF | Other involvements | Biology | ECA | Treatment | Outcome |
|------|--------------------------|-----|----------------------|-------------------|---------|-----|-----------|---------|
| 1    | 29                       | F   | Revealing            | Left              | Mediastinal lymph nodes | Hypercalciuria/ Lymphopenia | Nil | Oral CT (0,7 mg/kg/day) Methotrexate | Decrease of FP |
| 2    | 52                       | F   | Revealing            | Left              | Granulomatous anterior uveitis | - | Oral CT (1mg/kg/day) | Decrease of FP |
| 3    | 62                       | M   | Revealing            | Bending           | Mediastinal lymph nodes | Hypercalcemia | Nil | Oral CT (0.5mg/kg/day) | Decrease of FP |
| 4    | 45                       | F   | Revealing            | Left              | Bilateral granulomatous panuveitis | Lymphopenia | Oral CT (1mg/kg/day) | Decrease of FP |
| 5    | 40                       | F   | Revealing            | Bending           | Bilateral granulomatous anterior uveitis | Lymphopenia | Oral CT (1mg/kg/day) | Recurrence of FP |
| 6    | 39                       | M   | Left                 |                  | Bilateral granulomatous panuveitis | Hypercalcemia | Hypercalciuria | Nil | Oral CT (1mg/kg/day) | Decrease of FP |
| 7    | 50                       | M   | Revealing            | Bending           | Parotiditis | Lymphopenia | Oral CT (1mg/kg/day) | Decrease of FP |

Abreviations: ECA: FP : Facial Paralysis; F: Female; M : Male; CT : Corticosteroids
The treatment is mainly based on corticosteroid therapy (starting dose between 0.5 and 1 mg/kg/day) whose purpose is to reduce granulomatous in inflammatory lesions. Intravenous methyl prednisolone bolus may be prescribed in severe cases. This treatment should be initiated as early as possible to limit the risk of developing hemifacial spasm. The therapeutic response should be evaluated after 1 to 3 months. Treatment should be continued for at least 12 months [14]. In our series all patients received corticosteroid therapy at a dose of 0.5 to 1 mg/kg/day depending on the severity of the impairment and the presence of other serious systemic impairment corticosteroids usually yallow rapid clinical improvement. Only one case of recurrence was noted despite treatment. In our series, the evolution was favorable with an average time of 3 months the other immuno suppressants (cyclophosphamide, methotrexate, azathioprine, ciclosporine) are prescribed in case of failure of corticosteroids, corticodependence or particularly severe forms [15]. Rehabilitation appears essential for severe forms, in order to limits pasticse quelae (about 5 to 10% of cases despite well-conducted medical treatment). It is based in the initial phase on a work of symmetrization of the face to avoid the hyperactivity frequently observed on the healthy side [16]. The prognosis is essentially functional. It is imperative to look for an ocular complication including keratitis is secondary to corneal exposure (absence of occlusion, decreased tear secretion, associated trigeminal nerve involvement) and requiring urgent management [16]. The use of complementary explorations will be justified only in atypical or severe forms.

Conclusion

FP is rare during sarcoidosis. It is more common in women and would be most often revealing and left seat. The response to corticosteroids is rapid. Hence the interest of early diagnosis and early management. The prognosis is functional and is usually good.

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