



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 a 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 paralysis, 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 peripheral involvement were ruled 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 a 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: Mediastinal lymph node involvement in four cases, interstitial pneumopathy in four cases, granulomatous anterior uveitis in three cases, panuveitis in two cases, central neurological involvement in 3 two cases, meningeal involvement in a case, parotiditis in 1 case and erythema nodosum 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 caseous necrosis in 3 cases. Cerebral MRI showed a hypersignal of the white substance supra-tentorial non-specific in one case and a brain damage leptomenia under tentorial and contrast taking shooting of cerebellar and frontal pontocerebellar angles in another case. Oral corticosteroids allowed rapid regression of FP in six cases and this within an

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

Case	Age at the diagnosis of PF	Sex	Characteristics of PF	Other involments	Biology	ECA	Treatment	Outcome
1	29	F	Revealing Left	Mediastinal lymph nodes Interstitial pneumopathy Central neurological impairment	Hypercalciurae/ Lymphopenia	NI	Oral CT (0,7 mg/kg/day) Methotrexate	Decrease of FP
2	52	F	Revealing Left	Granulomatous anterior uveitis Central neurological impairment Interstitial pneumopathy	-		Oral CT (1mg/kg/day)	Decrease of FP
3	62	F	Revealing Bending	Mediastinal lymph nodes Interstitial pneumopathy	Hypercalcemia	NI	Oral CT (0,5mg/kg/day)	Decrease of FP
4	45	F	Revealing Left	Bilateral granulomatous panuveitis Mediastinal lymph node Meningitidis	Lymphopenia		Oral CT (1mg/kg/day)	Decrease of FP
5	40	F	Revealing Bending Recurrent	Bilateral granulomatous anterioruveitis Mediastinal lymph node Interstitial pneumopathy	Lymphopenia		Oral CT (1mg/kg/day)	Recurrence of FP
6	39	M	Left	Bilateral granulomatous panuveitis Central neurological impairment	Hypercalcemia Hypercalciuria	NI	Oral CT (1mg/kg/day)	Decrease of FP
7	50	M	Revealing Bending Recurrent	Parotidis Erythemanodosum Granulomatous anterior uveitis	Lymphopenia		Oral CT (1mg/kg/day)	Decrease of FP

Abreviations: ECA: FP : Facial Paralysis; F: Female; M : Male; CT : Corticosteroids

average of 3 months. One patient had a recurrence of the contralateral disease despite treatment. The clinical, para-clinical and evolutionary characteristics of the seven patients were reported in (Table 1).

Discussion

Our study describes the clinical, para-clinical 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 mean age of sarcoidosis patients is generally between 20 and 40 years with a second peak in frequency between 45 and 65 years [11]. The mean age 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 our series. Indeed, the FP had revealed sarcoidosis in six cases. 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 mphpnode involvement, and ocular involvement. The facial nerve involvement can fall into Heerfordt syndrome combining peripheral facial paralysis, bilateral anterior uveitis, bilateral parotiditis and fever. During this syndrome, the FP is very common it is almost of tenuni lateral and complete; it is sometimes bilateral. FP is often attributed to concomitant parotidite compression.

However, it does not always evolve in parallel with this one [13]. The FP. can sometimes precede parotiditis, as is the case with our 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 be neuritis of the face. Heerfordt syndrome has some similarities with sarcoidosis, of which it is only a particular form; Reticulo-endotheliosis 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 eliminating the etiologies. Idiopathic or refractory 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 chronic lymphadenopathy, other examinations will be performed mainly.

The treatment is mainly based on corticosteroid therapy (starting dose between 0.5 and 1 mg/kg/day) whose purpose is to reduce granulomatous 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. In case of favourable evolution the dose is gradually reduced. 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 1mg/kg/day depending on the severity of the impairment and the presence of other serious systemic impairment corticosteroids usually allow rapid clinical improvement. Only one case of recurrence was noted despite treatment. In our series, the evolution was favorable with in an average time of 3 months the other immunosuppressants (cyclophosphamide, methotrexate, azathioprine, ciclosporine) are prescribed in case of failure of corticosteroids, corticosteroid dependence or particularly severe forms [15]. Rehabilitation appears essential for severe forms, in order to limit postictic sequelae (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 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 sided. The response to corticosteroids is rapid. Hence the interest of early diagnosis and early management. The prognosis is functional and is usually good.

References

1. Ferriby D, Seze De J, Stojkovic T, et al. (2009) Long-term follow-up of neurosarcoidosis. *Neurology* 57: 927-929.
2. Pawate S, Moses H, Sriram S (2009) Presentations and outcomes of neurosarcoidosis: A study of 54 cases. *QJM* 102: 449-460.
3. Scott TF, Yandora K, Valeri A, et al. (2007) Aggressive therapy for neurosarcoidosis: Long term follow-up of 48 treated patients. *Arch Neurol* 64: 691-696.
4. Ben Ghorbel I, Bel Feki N, Ghoul El F, et al. (2012) Profil épidémiologique, clinique, paraclinique et évolutif d'une série monocentrique de sarcoïdose systémique. À propos de 90 observations. *Revue de Médecine Interne* 33: 167-168.
5. Essid A, Ben Fredj F, Abdelghani A, et al. (2009) La sarcoïdose systémique: A propos de 34 observations tunisiennes. *La Revue de Médecine Interne* 30: 117.
6. Nozaki K, Judson MA (2012) Neurosarcoidosis: Clinical manifestations, diagnosis and treatment. *Presse Med* 41: e331-e348.
7. Chapelon Abric C (2011) Localisations extrathoraciques graves de la sarcoïdose. *La Rev Med Interne* 32: 80-85.
8. Dubas F, Nicolas G (2001) Neurosarcoïdose. *Encycl Med Chir Neurologie* 17: 168.
9. Nowak DA, Widenka DC (2001) Neurosarcoidosis: A review of its intracranial manifestation. *J Neurol* 248: 363-372.
10. Colover J (1948) Sarcoidosis with involvement of the nervous system. *Brain* 71: 451-475.
11. Chapelon Abric C (2004) Épidémiologie de la sarcoïdose et ses facteurs de risque génétiques et environnementaux. *La Revue de Médecine Interne* 25: 494-500.
12. Joseph FG, Scolding NJ (2009) Neurosarcoidosis: A study of 30 new cases. *J Neurol Neurosurg Psychiatry* 80: 297-304.
13. Lambert V, Richards SH (1964) Facial Palsy in Heerfordt's Syndrome. *The Journal of Laryngology & Otology* 78: 684-693.
14. Stern BJ (2004) Neurological complications of sarcoidosis. *Curr Opin Neurol* 17: 311-316.
15. Doty JD, Mazur JE, Judson MA (2003) Treatment of corticosteroid-resistant neurosarcoidosis with a short-course cyclophosphamide regimen. *Chest* 124: 2023-2026.
16. Tankéré F, Bernat I (2009) Paralysie faciale à frigore: De l'étiologie virale à la réalité diagnostique. *La Revue de Médecine Interne* 30: 769-775.

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