

extraocular muscles, with intracranial extension and involvement of the cavernous sinus (Fig. 1a and c). CT scan of the chest showed bilateral hilar lymphadenopathy and small, well-defined lung nodes. An ^{18}F fluorodeoxyglucose PET/CT scan (Fig. 1b and d) showed normal uptake in the right orbit and the lymph nodes of the chest. An endoscopic biopsy of the endonasal cavernous sinus was performed, in which confluent, non-necrotising granulomas suggestive of sarcoidosis were found. Tuberculosis screening was negative.

Given these findings we started treatment with glucocorticoids, first 3 boluses of intravenous methylprednisolone from 1 g/day and then prednisone at a tapering dose of 1 mg/kg. There was rapid clinical improvement, although the orbital extension and established neurological damage showed less response, demonstrated on imaging tests. Given the response to steroids, immunosuppressants were not started.

In our patient, the orbital MRI findings suggested THS. After systemic examination and biopsy, we considered an unusual initial presentation of systemic sarcoidosis with major neurological symptoms. Isolated neurosarcoidosis is rare, since over 90% of patients also have sarcoidosis in other organs, especially the lungs and mediastinal lymph nodes.³ Cranial neuropathy is the most common manifestation of neurosarcoidosis.² Diagnosing neurosarcoidosis is often difficult, because the clinical manifestations and findings of imaging studies can be mimicked by several other diseases. Brain MRI is the most sensitive diagnostic imaging test.³

This case highlights that sarcoidosis can present in unusual ways, masking neurological disorders.

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Response to: Current Status of Treatment With Intra-Articular Infiltrations in Juvenile Idiopathic Arthritis[☆]



Respuesta a: Estado actual del tratamiento con infiltraciones intra-articulares en la artritis idiopática juvenil

Dear Editor,

We read the publication by Nieto-González and Monteagudo¹ in *REUMATOLOGÍA CLÍNICA* with interest, where a narrative review is undertaken of the literature in connection with the practice of using intra-articular infiltrations with corticoids (IAIC) in patients with juvenile idiopathic arthritis (JIA). We would like to describe the results of a survey of all of the members of the Spanish Paediatric Rheumatology Society (SERPE) in 2017, in which they were asked about their habitual practice in connection with several aspects of the said technique.

The survey consisted of 10 questions devised by the Scientific Committee of the 2017 National Congress of the said society. The questions refer to the medical speciality of the staff who perform IAIC (rheumatologist, paediatrician, rehabilitation doctor or orthopaedic surgeon); the type of JIA used in the said procedure; indication of analgesia and/or sedation during the process; drug(s) used; details of the IAIC technique (asepsis, washing in saline solution, dilution of the corticoid and others); maximum number of joints infiltrated in a single session; maximum number of IAIC in the same joint in one year; recommendations after infiltration; complications after infiltration; and differences between children according to their age. The “SurveyMonkey”

platform was used, which makes it possible to create online surveys, (<https://es.surveymonkey.com>). This platform interprets the replies to surveys and creates basic descriptive statistics for them.

Eighty-five of the 120 members contacted replied. The survey results showed the heterogeneity of the range of professionals who perform IAIC for JIA, as other authors have reported in the past.² The appended table shows the said results. In general, the answers to the survey reflect the absence of recognised treatment guides for this technique, showing the existence of practice that is “based on art” in the hospitals that treat young people and children with rheumatic diseases. A North American study that was also based on a survey reached similar conclusions.³

This study has several limitations, the chief one of which may be the fact that only 85 or 70% of 120 members responded to the survey. Additionally, not all of the doctors who treat rheumatic diseases in children in our country belong to the SERPE, although the majority do. On the other hand, one potentially interesting analysis was not carried out: to evaluate whether, within the heterogeneous nature of the replies, they could be found to be more homogeneous if classified according to the speciality of the respondent (rheumatologist, paediatrician, rehabilitator or orthopaedic surgeon).

There are other relevant aspects in connection with IAIC technique which we believe were included in the survey. One of these is the increasing importance of ultrasound scan imaging in paediatric rheumatology, as in recent years it has come to be included as another tool⁴ for use when infiltrating several locations that are considered to be “difficult” (the temporomandibular joint,⁵ tenosynovitis,⁶ the tarsal joint⁷). On the other hand, in all fields of paediatrics sedation-analgesic techniques are being adopted to improve the quality of care for paediatric patients; including IAIC, according to several recent publications.^{8–10}

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Table 1
Results of the survey of 120 members of the SERPE with replies from 85 of them

Questions	Never	Sometimes	Often	Always
<i>Who performs IAIC in your hospital?</i>				
Orthopaedic surgeon: 11%	We do not perform IAIC: 1.2%			
Paediatrician: 87%	Rehabilitation doctor: 11%			
Rheumatologist: 19%				
<i>In what clinical situations do you perform IAIC?</i>				
Monoarthritis, %	2,3	1,2	35	62
Oligoarthritis, %	0	18	71	11
Polyarthritis, %	2,4	47	45	6
Relapse, %	1,2	27	61	11
<i>What about analgesia and sedation?</i>				
Chlorethyl spray, %	60	17	13	10
EMLA, %	27	25	25	22
Nitrous oxide, %	17	28	46	9
Benzodiazepines, %	28	42	28	2,6
Fentanyl, %	69	20	12	0
General anaesthetic, %	42	27	30	1,3
Local anaesthesia, %	45	30	12	12
<i>Which drug do you use?</i>				
Triamcinolone acetonide, %	3,6	17	35	45
Triamcinolone hexacetonide, %	31	37	22	9
Betamethasone, %	75	15	10	0
Hyaluronic acid, %	68	29	3	0
Itrium, %	64	35	0	0
<i>What dose do you use depending on joint size?</i>				
0,5 mg/kg in small joints and 1 mg/kg in large joints: 50%				
20 mg in small joints and 40 mg in large joints: 50%				
<i>In respect of the technique</i>				
Guided by ultrasound scan, %	13	50	31	5
Sterile gloves WITH surgical field, %	6	8	13	72
Sterile gloves WITHOUT surgical field, %	53	17	16	14
Only skin sterilisation, %	89	4	5	2,5
Dose calculated according to weight, %	23	34	19	23
I consider the total dose, %	14	18	25	4
I obtain informed consent, %	15	13	18	54%
<i>How many joints do you infiltrate in the same session?</i>				
	Two	Three	Four	More than four
Do you think that the same joint should not be infiltrated > 3 times per year?	30%	31%	35%	4%
<i>In connection with joint locations</i>	Yes: 76%		No: 24%	
I infiltrate for tenosynovitis		Yes: 63%	No: 33%	
I infiltrate the tarsal joint		Yes: 61%	No: 33%	
I infiltrate TMJ		Yes: 30%	No: 70%	
I infiltrate for IFP, MCF		Yes: 71%	No: 29%	
I infiltrate for dactylitis		Yes: 50%	No: 50%	
I infiltrate the hips		Yes: 60%	No: 40%	
I infiltrate the sacroiliac		Yes: 6%	No: 94%	
<i>In post-filtration recommendations</i>				
I recommend normal life without excessive effort, %	28	25	23	23
I recommend rest for 12-24 hrs., %	7	7	13	73
I recommend oral analgesia, %	14	41	24	21
I make an appointment for them in 1-2 weeks for evaluation, %	0	22	20	58
I do not evaluate again, they contact in case of no improvement, %	74	14	7	5
<i>Have you seen these complications?</i>				
	Never	Rarely (< 1/100)	Often (1/10-1/100)	
Cutaneous atrophy, %	8	64	27	
Septic arthritis, %	95	4	0	
Vasovagal reaction, %	55	41	3,5	
Haemorrhage due to arterial puncture, %	90	9,4	0	
<i>Do you establish differences for the smallest patients (< 5 years)?</i>				
	Yes	No		
I use ultrasound scan more often, %	49	51		
I use more sedation/analgesia methods, %	95	5		
I reduce the maximum number of IAIC, %	35	64		
I use a lower dose of steroid in large joints, %	63	37		

EMLA: Eutectic Mixture of Local Anaesthetic; IAIC: intra-articular infiltration with corticoids; IFP: proximal interphalangeal; MCF: metacarpophalangeal; SERPE: Sociedad Española de Reumatología Pediátrica; TMJ: temporomandibular joint.

Although IAIC is not a technique that involves important adverse events and does not require long training to be performed in a sufficiently expert way, the authors believe it necessary to prepare guides or recommendations for the technique, for paediatric and young patients (Table 1).

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Paravertebral pseudotumour in granulomatosis with polyangiitis[☆]



Seudotumor paravertebral en granulomatosis con poliangeítis

Dear Editor,

Granulomatosis with polyangiitis (GPA) is a type of vasculitis associated with neutrophil anti-cytoplasm antibodies (ANCA) which may affect small and medium calibre vessels.¹ It is characterised by granulomatose lesions and/or necrosing vasculitis in the upper and/or lower respiratory tract, glomerulonephritis and, less often, mucocutaneous, orbital and neurological involvement, among others.¹ Pre- or paravertebral pseudotumour lesions are rare, and there are few reports of them in GPA.^{2,3} A case is presented of paravertebral pseudotumour as an incidental finding in the context of systemic GPA involvement.

A 60 year-old man with arterial hypertension and chronic kidney disease that had evolved over 2 years, requiring haemodialysis and previous renal biopsy with sclerosed glomeruli, pericapsular and interstitial fibrosis and atrophic tubules. He visited due to weight loss over 3 months and palpable purpura in the lower limbs. He had symptoms of a compromised upper airway, renal involvement with haematuria and proteinuria in a non-nephrotic range with deterioration of kidney function. Laboratory tests found normocytic normochromic anaemia (haemoglobin 7.1 g/dl and haematocrit 21.9%), normal white blood cells (5,600/mm³), accelerated erythrocytation (70 mm/hr.), raised reactive C protein (3.19 mg/dl), creatinine (6.3 mg/dl), proteinuria/creatininuria quotient 2.1 and macroscopic hematuria. ANCA were positive, PR3 ELISA and cytoplasmic pattern in immunofluorescence. The antinuclear antibodies, anti-Ro/SS-A, anti-La/SS-B, anti-DNA and viral serologies for hepatitis B, C and HIV were negative. Computed

tomography (CT) of the paranasal sinuses showed thickening of the mucosa and bone erosion, and high resolution CT of the thorax revealed centrolobulillar nodules and a paravertebral pseudotumour lesion (Fig. 1A). CT guided biopsy was performed and showed findings that could be associated with vasculitis without granulomatosis (Fig. 1B and C).

Due to the said clinical findings, the positive ANCA and the histological changes in the paravertebral mass, GPA was diagnosed and induction therapy commenced with pulses of methylprednisolone and cyclophosphamide (15 mg/kg/every 15 days/6 doses). Thoracic CT was performed after 4 months, finding a significant reduction in the size of the lesion (Fig. 1D).

Infiltrating pseudotumour lesions are frequent in patients with GPA. They may be located in the orbit, paranasal sinuses, the mediastinum, breast, kidney and retroperitoneum, among other locations.¹ Paravertebral lesions are rare,^{2,3} and to our knowledge only 10 cases have been reported in this location, although there are other reports of mediastinal lesions.^{2,3} They may present at the moment the disease starts or during its evolution.^{2,3} The frequency of such lesions may be underestimated as 50% of cases are asymptomatic and are not associated with bone erosion or the compression of adjacent structures.² Some patients may experience chronic thoracic pain.³ Thoracic CT with contrast and magnetic resonance imaging studies are the main studies suggested for the detection of these lesions and to establish differential diagnoses. Histopathology is essential for diagnosis.²⁻⁴ It also makes it possible to rule out malignancy and other differential diagnoses such as tuberculosis, mycosis, sarcoidosis, histiocytosis and disease associated with IgG4.⁴⁻⁶ This lesion location gives rise to difficulties in obtaining histological samples, so that it is suggested that a CT-guided biopsy be performed.^{2,3} The samples obtained usually show granulomatose infiltrates, rarely vasculitis and calcifications.²⁻⁷ After treatment, the inflammatory lesions may reduce in size, while the lesions that have evolved the most and fibrosis are unchanged.²⁻⁶

To conclude, GPA should be suspected if there is a finding of paravertebral pseudotumours within an appropriate clinical context.

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