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SCIENTIFIC LETTER

Idiopathic facial aseptic granuloma: Clinical, pathological, and ultrasound characteristics☆

Granuloma aséptico facial idiopático: características clinicopatológicas y ecográficas en 7 casos

Dear Editor,

Idiopathic facial aseptic granuloma (IFAG) is a recently described disease that affects the paediatric age group.[1] It is characterised by the development of one or more nodules in the cheeks and the differential diagnosis must include other facial nodules acquired in childhood, including pilomatrixomas, dermoid cysts, bacterial infection by *Leishmania* or mycobacteria and vascular malformations. For this reason, it is probably underdiagnosed.

We conducted a retrospective observational study of all cases of IFAG diagnosed in our hospital over a 12-year period. Table 1 summarises the characteristics of these cases. The mean age of the patients was 3 years. Three of the patients had multiple lesions. Five of the patients received oral antibiotherapy: 2 amoxicillin–clavulanic acid, 1 cefuroxime, 1 clarithromycin and 1 amoxicillin–clavulanic acid followed by azithromycin with no clear evidence of improvement. The disease tended to resolve spontaneously by a mean of 13 months. Two patients underwent surgical removal of the lesions. None of the patients experienced complications or recurrence during the follow-up.

The histological examination revealed a granulomatous reaction of the dermis, with lymphocyte and plasma cells and negative results of staining for detection of microorganisms (Fig. 1C). In one of the cases, the examination found remnants of an epidermoid cyst.

The ultrasound scan revealed hypoechoic oval structures in the dermis with the longest diameter parallel to the skin surface. There were no calcium deposits. The posterior edge was even except in 1 case in which it exhibited mildly rolled edges (Fig. 1D). The posterior hypodermis was hyperechoic. The presence of peripheral fluid was variable, and there was no fluid within the lesion in any case.

Idiopathic facial aseptic granuloma is characterised by the development of a painless, hard red or purple nodule in the cheek.[2] It has a characteristic location: in 2/3 of cases, it is located within a triangle with vertices at the earlobe, the external corner of the eye and the mouth corner. There is usually a single lesion, although some patients may have more than 1. It tends to regress spontaneously in a mean of 11 months.[2] All patients in our case series fit these clinical characteristics and course of disease.

The mean age at onset is 46 months, with a range of 8 months to 13 years.[1,2] It is more frequent in female patients, although this was not the case in our series.

The pathogenesis of IFAG has yet to be determined. One of the current hypotheses is that it could belong in the spectrum of granulomatous rosacea in children.[3] The concomitant presence of chalazions (commonly associated with rosacea), described in up to 2/3 of patients at the time of diagnosis of IFAG, the subsequent development of rosacea lesions in a large number of patients, the detection of perifollicular histocyte infiltration in the histological examination, typical of granulomatous rosacea, or the response to antibiotic agents used for treatment of rosacea are some of the features that suggest an association with granulomatous rosacea.[3–5] In our case series, one patient developed perioral rosacea papules and another patient had a chalazion.

The possibility of an infectious aetiology seems to have been excluded, at least when it comes to known pathogens.[2]

A third hypothesis suggests the development of a granulomatous reaction in response to an embryonal remnant.[2] This possibility is supported by a case where the histological examination revealed remnants of an epidermoid cyst,[2] which we also found in one of our patients.

Lastly, other proposed aetiologies include a persisting reaction to an insect bite or traumatic injury.

The histological presentation of IFAG is characterised by a cutaneous inflammatory granulomatous infiltrate composed mainly of lymphocytes, histocytes, neutrophils and multinucleated foreign body giant cells in the absence of calcification or ghost cells.[2]

The ultrasound examination of skin lesions reveals hypoechoic oval structures in the dermis with posterior hyperechogenicity with no calcification.[2,4] Patients with acute inflammation present with perilesional fluid, which disappears as the lesion resolves. In most cases of IFAG, there is no fluid within the lesion.[6]

When it comes to treatment, the previous literature describes improvement with long courses (2–3 months) of oral clarithromycin or doxycycline and topical metronidazole or ivermectin.[3,5] Amoxicillin–clavulanic acid, azithromycin, erythromycin and cefalexin have proven ineffective.

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Table 1 Patient characteristics.

Case	Sex	Age	Clinical features	Histological examination	Ultrasound	Time to resolution
1	Male	5 years	Indurated nodule in right cheek	Yes	Yes	Surgical removal
2	Male	4 months	Several papules in left cheek. Plaque with central crust	Yes	No	15 months
3	Male	9 years	Soft nodule in left cheek	Yes	Yes	15 months
4	Male	2 years	Nodule in left cheek Perioral pink papules Episodes of facial erythema	Yes	No	9 months
5	Male	8 months	Purulent papule in left cheek	No	No	14 months
6	Female	2 years	Nodule in right cheek Bleeding Papule in left cheek Chalazion	Yes	Yes	Surgical removal
7	Female	6 years	Nodule in left cheek	No	Yes	Ongoing

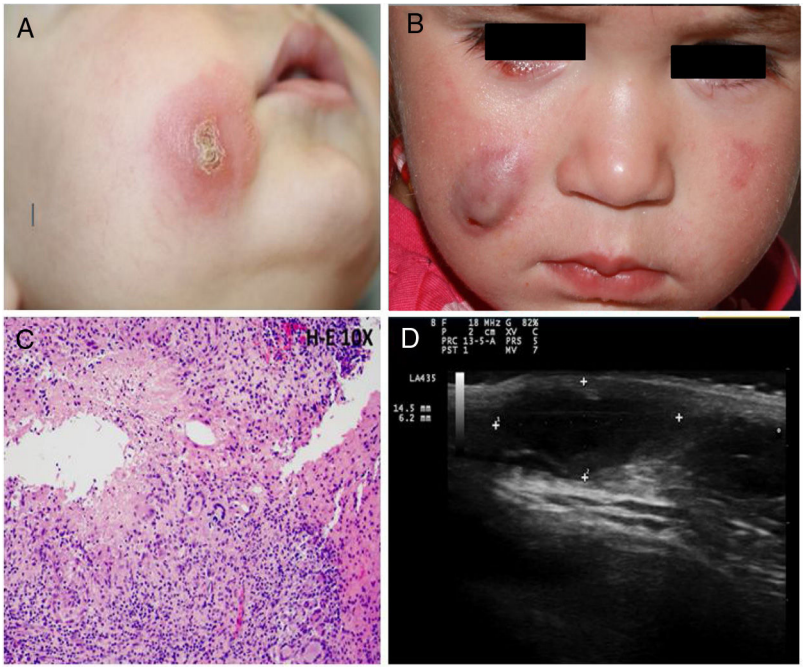

Figure 1 (A) Clinical image of case 2. (B) Clinical image of case 6 showing a chalazion in the right lower eyelid. (C) Histological examination of the lesion in the right cheek of case 6, showing a cutaneous infiltrate of lymphocytes and plasma cells with granulomas composed of histocytes and multinucleate giant cells. (D) Ultrasound of lesion in the right cheek (case 6) showing a hypoechoic structure with a rolled posterior edge.

The aim of this study was to bring attention to a disease that is probably underdiagnosed. Since this disease resolves spontaneously and is located in an area that has an aesthetic impact, the use of invasive diagnostic tests must be justified, and ultrasonography is an important non-invasive tool in its differential diagnosis. In the future, additional descriptions of IFAG cases will probably help elucidate the aetiology and optimal management of this disease.

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Controlled asystole donation in the paediatric patient☆

Donación en asistolia controlada en el paciente pediátrico

Dear Editor:

Despite the high number of organ transplantations performed in countries like Spain, the availability of organs for paediatric transplantation continues to be limited, which leads to higher morbidity and mortality in this population while waiting.

Donation protocols traditionally involve donation after the diagnosis of brain death in the donor (heart-beating donation), but in recent years protocols have also been developed for non-heart-beating donation, or donation after circulatory death (DCD). Donation after circulatory death is categorised based on the revised Maastricht classification (Paris 2013) into uncontrolled DCD, category I (dead on arrival, or sudden cardiac arrest without performance of cardiopulmonary resuscitation) and category II (sudden cardiac arrest with unsuccessful resuscitation) and controlled DCD, category III (awaiting circulatory death after withdrawal of life-sustaining therapies) and category IV (cardiac arrest in a brain-dead donor).[1,2]

Category III includes patients whose condition has led to the decision of withdrawal or withholding of life-sustaining care. Following this decision, it is considered good clinical practice to consider the patient as a potential organ and tissue donor. The decision to withhold life-sustaining treatment should be made before, separately and completely independently from potential decisions regarding donation after death and the donation process. The transplant coordination team has to assess the appropriateness of the candidate and ensure that the time expected to elapse from withdrawal of life-sustaining treatment to death will be compatible with organ donation and not exceed the warm ischaemia time threshold established by the transplantation care team. The paediatric intensive care team is responsible for the patient and, completely removed from the donation process, for providing end-of-life care to ensure the well-being and comfort of the patient and for withdrawing life-sustaining therapies. This team is also responsible for death certification, which according to current law, requires verifying the absence of spontaneous circulation and breathing for a period of at least 5 minutes.[2]

The outcomes of organ transplantation in this donation category, such as kidney and liver transplantation, have not been worse compared to the outcomes of transplants from heart-beating brain-dead donors.[1,3]

Category III controlled DCD has been performed successfully in adult intensive care units and currently amounts to 30% of all donations.[1,3,4] Although this type of donation has grown significantly in recent years in countries like the United States and Canada,[1,4] it continues to be rare in Spain.[5]

We present the case of a girl aged 15 months with non-compaction dilated cardiomyopathy and severe ventricular dysfunction who required support with an external ventricular assist device and was placed on the transplant waitlist. At 22 days from admission in the paediatric intensive care unit (PICU) she developed convulsive seizures, with imaging revealing the presence of a subdural haematoma with midline shift that required surgery. After 72 hours, there was evidence of an acute ischaemic stroke of the left middle cerebral artery that in 3 days had progressed to massive strokes with bilateral involvement of the anterior and middle cerebral arteries and the basal ganglia. Given the poor prognosis, the decision was made to withdraw life-sustaining treatment. The donation protocol was activated after this decision, and the patient was evaluated by the transplant coordination team, while the family expressed the wish to donate. The assessment by the transplant team found positive results for the kidney, liver (although a compatible recipient was not found for this organ) and tissues. The patient had not gone through brain death, so this was a controlled DCD donation. The process involved transport of the patient to an operating room (while the urology team got ready in an adjacent room), where mechanical ventilation and the ventricular assist device were withdrawn. During the entire process, the paediatric intensive care specialists in charge of the patient maintained sedation and analgesia per the life-sustaining therapy withdrawal protocol. Sixteen minutes after supportive care was withdrawn, the patient

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