Hepatic vasculitis mimicking multiple liver abscesses in Cogan's Syndrome

Dinis de Freitas J[1], Costa F[1], Rovisco J[1]

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To the Editor,

Cogan's syndrome (CS) is a rare chronic inflammatory disorder, classically characterized by inflammatory eye disease and vestibulo-auditory symptoms, but can also involve systemic vasculitis. It was first described by an ophthalmologist, David G. Cogan in 1945, as a non-syphilitic interstitial keratitis (IK), coexisting with Meniere-like vestibuloauditory symptoms[1,2]. We report a rare case of a patient with typical Cogan syndrome who developed hepatic vasculitis.

A 20-year-old female presented in the emergency department with rotational vertigo, tinittus and progressive hearing loss for 15 days, acute redness of eyes for 4 days and right flank pain with one-week progression. Redness of the eyes was associated with diminished vision and photophobia. Physical examination revealed circumciliary congestion of eyes with preserved visual acuity and bilateral sensorineural hearing loss. Pain in the right upper abdomen without peritoneal irritation was present. Besides low to moderate grade fever no other remarkable signs were present at physical examination. She was diagnosed with interstitial keratitis upon observation by Ophthalmologist. Significant findings in laboratory investigations included raised erythrocyte sedimentation rate 70 mm/hour and C-reactive protein 24 mg/dl. Hepatic liver enzymes and total bilirubin were also elevated, while the rest of the routine metabolic profile was normal. Antineutrophil cytoplasmic antibody, antinuclear antibodies and infectious serologies were negative. Pure tone audiometry showed bilateral sensorineural hearing loss. An abdominal computed tomography (CT) scan revealed multiple liver abscesses further characterized by magnetic resonance imaging (MRI) (Figure 1-A,B,C). A brain MRI revealed "enhancement of the VIII cranial pairs and the membranous labyrinths of the inner ear, bilaterally, aspects suggestive of neuritis and bilateral labyrinthitis" (Figure 1-D,E). The remaining evaluation was unremarkable (blood cultures, chest x-ray, echocardiogram, lumbar puncture and urinalysis). Our patient started 1.5 mg/kg/day of prednisone. Abscesses and red eye progressively resolved and inflammatory markers normalized. Neurossensorial deafness was not reversible. Corticosteroids were gradually tapered in the next 6 months and for the past 2 years the patient has been in remission without immunosuppression medication.

In this case the diagnosis of typical CS was established since vestibular symptoms and KI manifested almost simultaneous contrary to atypical CS where a delay of more than 2 years between symptoms occur[2,3]. Although we lack histologic confirmation, hepatic vasculitis mimicking abscess, were assumed to be a sys-

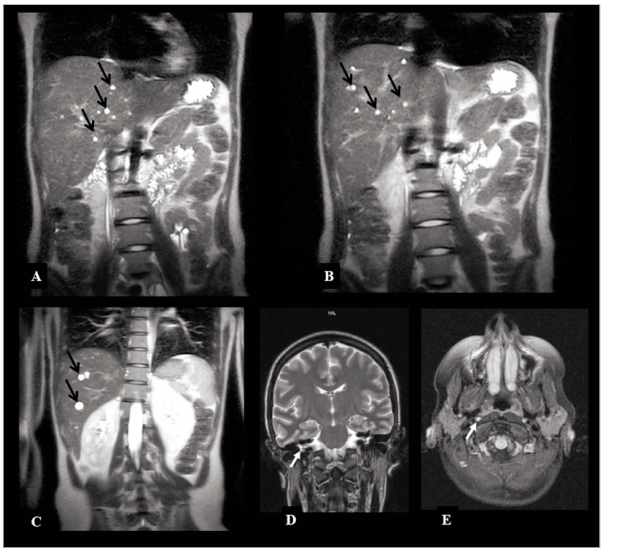

FIGURE 1. A-C: Black arrows highlight multiple nodules appearing hyperintense in T2W1 MRI sequence suggesting hepatic abscesses;
D-E: Brain MRI reveals enhancement of the VIII cranial pairs (white arrows), as well enhancement of the membranous labyrinths of the inner ear, bilaterally in T2, aspects suggestive of neuritis and bilateral labyrinthitis.

1, Reumatologia, Centro Hospitalar e Universitário De Coimbra, E.P.E.

temic manifestation of CS, as septic screening was unremarkable for infection.

In a literature review, by Grasland *et al.*, systemic vasculitis was present in 22% of the 99 cases of atypical CS identified. Those with systemic vasculitis had a more aggressive disease and a worse outcome[4]. Although there is usually large and/or medium vessel commitment, any size vessel may be affected[5]. Gastrointestinal involvement is common and is present in a quarter of the patients. It can be rectal bleeding, abdominal pain and hepatomegaly[6]. In a 32 case series from 2004, only two cases of hepatic vasculitis were reported[4]. Corticosteroids are the first line of treatment, although there are no definitive therapeutic recommendations, as no controlled trials have been made[7]. Patients without systemic disease generally have a good prognosis. Those who develop serious vasculitis, have an increased risk of death due to complications[4]. Our patient presented with a severe systemic vasculitis and responded favorably to the timely institution of steroid therapy with complete resolution of the aseptic abscesses.

This case shows the importance of a high index of suspicion to effectively manage CS, as prompt diagnosis and initiation of treatment is needed to prevent life-threatening complications.

CORRESPONDENCE TO
João Pedro Dinis de Freitas
Reumatologia, Centro Hospitalar e Universitário De Coimbra
Praceta, R. Prof. Mota Pinto
3075, Coimbra
E-mail: pedrodinisfreitas@gmail.com

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