Clinical Rheumatology
https://doi.org/10.1007/s10067-020-05538-x

CLINICAL IMAGE

Pediatric inflammatory multisystem syndrome (PIMS) presenting with retropharyngeal phlegmon mimicking Kawasaki disease

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Received: 6 November 2020 / Revised: 1 December 2020 / Accepted: 5 December 2020

There have been several recent reports of children presenting with severe multisystem inflammatory syndrome (PIMS) resembling complete or incomplete Kawasaki disease (KD) during COVID19 pandemic. Being a novel disease, limited data is available for its identification and management. We report a child presenting with fever, neck swelling, and generalized edema, who was diagnosed as PIMS. MRI neck showed retropharyngeal phlegmon resembling similar finding in KD.

Presentation

A 4-year-old boy presented with high fever for 6 days along with a painful neck swelling associated with bilateral nonpurulent conjunctivitis, facial puffiness, and extreme irritability. He had tachycardia, hypotension, S3 gallop, and generalized edema. The neck swelling was extremely painful causing neck stiffness. With a working diagnosis of sepsis and myocarditis empirical broad spectrum, antibiotics were started, but a possibility of KD and PIMS was also considered. Echocardiography showed grade 1 systolic dysfunction, and MRI neck (Fig.1) demonstrated a retropharyngeal fluid collection with soft tissue edema, likely to be a retropharyngeal phlegmon, as described in KD. COVID RT-PCR was negative but COVID IgG was positive.

He was treated with IVIg @2 g/kg followed by IV methyl prednisolone @5 mg/kg/day for 3 days. Fever subsided within 48 h of IVIg, and the inflammatory markers started normalizing (Table 1) on repeat testing after3 days of methylprednisolone. The child was discharged on low-dose aspirin and oral prednisolone 2 mg/kg/day to be tapered over 15 days, with advice for a follow-up echocardiography after 14 days. The child is doing well at present, and repeat echocardiography is within normal limit.

Discussion

There has been several case reports of KD presenting with retropharyngeal edema/phlegmon [1, 2]. This fact that cervical lymphadenopathy in KD can be associated with deeper soft

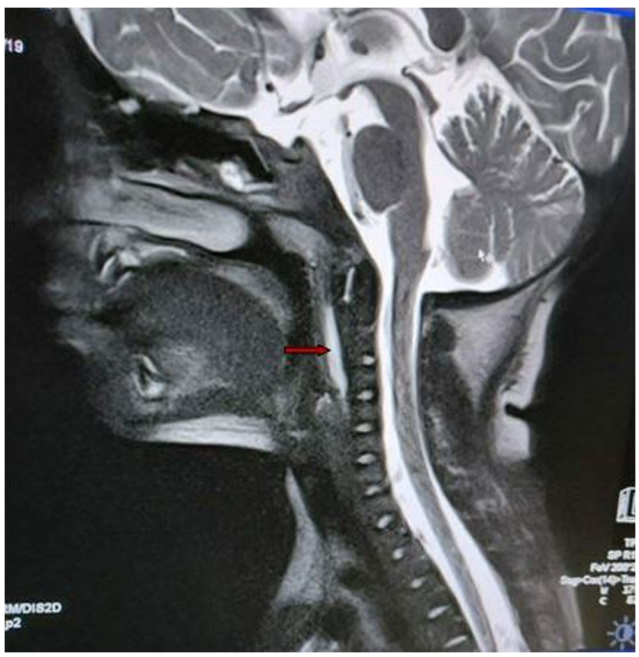

Fig. 1 MRI neck showing linear retropharyngeal fluid collection with soft tissue edema without any compression over the airway

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Published online: 13 January 2021

Table 1 Comparison between the different laboratory markers before and after starting treatment with IVIg and methyl prednisolone, respectively

		On admission (D1)	Day of starting IVIg (D2)	After 48 h of IVIg completion (methyl prednisolone initiated)	Before discharge
Investigations	Normal range	22/09/2020	23/09/2020	25/09/2020	29/09/2020
Total leucocyte count (/cmm)	4000–10,000	9600	9410	9910	8420
Differential count (%) N–neutrophil L-lymphocyte	N 40–60 L 20–40	N 66 L 29	N 79 L 18	N 66 L 29	N 34 L 58
Hemoglobin(g m%)	11–14	9.4	10.2	9.1	10.6
Platelets (× 109/l)	150–400	218	103	319	617
CRP (mg/l)	< 5	318.3	191	94.1	30
Urea (mg/dl)	7–20	18	17	17	18
Creatinine(mg/dl)	0.3–0.8	0.39	0.38	0.36	0.36
Sodium (mEq/l)	135–145	128	134	137	136
Potassium (mEq/l)	3.5–5	3.2	3.5	4.4	4
Total Bilirubin (mg/dl)	0.1–1.2	0.39	0.4	0.4	0.38
Total protein (g/dl)	6–8	4.4	6.4	6.7	7.2
Albumin(g/dl)	3.5–5.5	2.5	3.2	3.5	4
ALT (U/l)	7–50	10	12	13	12
AST (U/l)	10–40	32	34	33	34
Ferritin (ng/ml)	< 250	570.2	340.5	198	76
Interleukin 6	< 7 pg/ml	91.2	57	12	12
NTpro BNP	< 450 pg/ml	29,562	12,764	5436	2917

tissue inflammation leading to non-suppurative edema is being increasingly recognized and has been included as "Other Clinical Finding" in the AHA 2017 Scientific Statement on KD [3].

With the onset of the COVID 19 pandemic, cases presenting with multisystem hyperinflammation (PIMS) resembling atypical KD are being frequently reported globally [4, 5]. Though cervical lymphadenopathy is present as an associated finding in some of these patients, the authors did not come across any description of retropharyngeal edema in the available literature. This child presented with high fever with painful torticollis and had other features of systemic inflammation with myocarditis resembling PIMS. Although there was no history of exposure to COVID, he tested positive for COVID IgG signifying an asymptomatic past infection. He responded very well to IVIg and IV methyl prednisolone with improvement of biochemical parameters and echocardiography findings.

Compliance with ethical standards

Disclosures None.

Patient consent Obtained. Written consent has been attached.

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