Multiple subarachnoid and bilateral stellate ganglion blocks in a child with newly diagnosed mixed connective tissue disease

Sir,

Mixed connective tissue disease (MCTD) is a chronic immune-mediated disease characterised by vasculitis, myositis, polyarthritis, pulmonary fibrosis, and Raynaud's phenomenon (RP).[1] We describe the management of severe pain and digital discolouration secondary to RP in all limbs in a child of MCTD.

A 12 year, 29 kg female, a diagnosed case of MCTD, on oral pentoxyphylline 250 mg, nifedipine 10 mg, prednisolone30mg, aspirin 37.5 mg, naproxen 250 mg, and hydroxychloroquine 100 mg, was referred to the pain clinic for the management of severe pain and discolouration of digits [Figure 1].

On examination, the child was afebrile with a heart rate of 104/min and a respiratory rate of 18/min. The patient had dry scaly skin; ulcers over elbows, ankles, and toes; discolouration of multiple digits with a high numerical pain rating scale (NRS) of 7–9 [Figure 1]. Oxygen saturation (SpO_2) was unrecordable in multiple digits[Table 1]. Blood investigations were normal except for a white blood cell count of 22,000/mm^3.

Figure 1: Discolouration of patient's digits in upper and lower limbs on various dates

The child was admitted to the paediatric ward. After parental informed consent, serial subarachnoid block (SAB) and ultrasound-guided stellate ganglion block (USGB) were undertaken with parental presence in the pain clinic. The intravenous cannula was secured and standard monitors attached. Right USGB was performed with 26G/1.5inches hypodermic needle with 5 ml ropivacaine (0.25%) and clonidine 10 µg, using a linear 5–13 MHz ultrasound (US) probe (Fujifilm SonoSite Edge). The block was performed in the long axis.The needle was inserted from the lateral to the medial side of the neck, placing the needle tip between the longus colli muscle and the carotid sheath at the level of the C7vertebra. Saline was injected to ensure the correct needle tip position followed by the drug.[2] After 30 minutes, SAB with bupivacaine (hyperbaric) 2.5 mg and clonidine 15 ug was performed with a Quincke needle (27G) at L4-L5 level. After 2 hours, left USGB was performed similarly. The child was monitored for another two hours before shifting to the ward. There was significant pain relief after the first set of blocks [Table 1]. A total of three sets of blocks were given on four successive days [Table 1]. The patient's clinical condition improved significantly and she was discharged on the fifth day [Figure 1, Table 1]. After a month, she presented with NRS 3 in the right upper limb with purulent discharge from the right foot, for which she was admitted. Right USGB was given on two alternate days which resulted in complete pain relief. Debridement of the infected gangrene in both the feet was done under spinal anaesthesia with 10 mg bupivacaine (hyperbaric) and 15 ug clonidine with a 27G spinal needle [Table 1]. The child was discharged after three days.She continues to be pain-free for the last 8 months. In a recent follow-up, she was found to have lost a few toes of the right foot by auto-amputation, though her feet, fingers, and hands have recovered [Figure 1].

Vasculopathy is the first feature of MCTD. RP secondary to vasculopathy can frequently occur and result in immense pain and discolouration of the extremity. Medical management may take 2–4 weeks for complete effect, during which vasculitis might progress with pain and auto-amputation of digits. Sympathetic blocks at this stage are crucial for treating pain and vasospasm.

In the present patient, bilateral stellate ganglion blocks (SGB) were planned under US guidance to minimise the risk of injury to adjacent nerves, plexus, and vessels in the neck. Bilateral USGB is scarcely

Table 1: Sequence of upper and lower limb blocks

Date	Right upper limb								Left upper limb							
	Baseline				Post USGB				Baseline				Post USGB			
	Digit	NRS	Colour	SpO_2	Digit	*NRS	Colour	SpO_2	Digit	NRS	Colour	SpO_2	Digit	*NRS	Colour	SpO_2
09/12/20	1,2	8-9	Blue	NR	1,2	3/8 h	Blue	NR	1,2	7-8	Blue	NR	1,2	3/8h	Blue	NR
	3,4,5	9	Blue	98%	3,4,5	3/8 h	Blue	98%	3,4,5	7	Blue	98%	3,4,5	3/8	Blue	98%
10/12/20	1,2	4	Blue	NR	1,2	1/20h	Blue	IW	1, 2	4	Blue	NR	1,2	1/20h	Blue	IW
	3,4,5	4	Blue	98%	3,4,5	1/20h	Blue	98%	3,4,5	4	Blue	98%	3,4,5	1/20h	Blue	98%
12/12/20	1,2	5	Blue	IW	1,2	1/36h	Blue	98%	1,2	5	Blue	IW	1,2	1/36h	Blue	98%
	3,4,5	5	Normal	98%	3,4,5	1/36h	Normal	98%	3,4,5	5	Normal	98%	3,4,5	1/36h	Normal	98%
Date	**Right lower limb**								**Left lower limb**							
	Baseline				Post Block				Baseline				Post Block			
	Digit	NRS	Colour	SpO_2	Digit	*NRS	Colour	SpO_2	Digit	NRS	Colour	SpO_2	Digit	*NRS	Colour	SpO_2
09/12/20	1,2,4,5	7	Black	NR	1,2,4,5	2/24h	Black	NR	1,2,5	7	Black	NR	1,2,5	2	Black	NR
	3	7	Blue	NR	3	2/24h	Blue	IW	3,4	7	Blue	NR	3,4	2	Blue	IW
10/12/20	1,2,4,5	3	Black	NR	1,2,4,5	1/24h	Black	NR	1,2,5	3	Black	NR	1,2,5	1/24h	Black	NR
	3	3	Blue	IW	3	1/24h	Blue	IW	3,4	3	Blue	IW	3,4	1/24h	Blue	IW
12/12/20	1,2,4,5	4	Black	NR	1,2,4,5	0/36h	Black	NR	1,2,5	4	Black	NR	1,2,5	0/36h	Blue	NR
	3	4	Blue	IW	3	0/36h	Blue	97%	3,4	4	Blue	IW	3,4	0/36h	Blue	97%
Discharged home																
Date	**Right upper limb**								**Left upper limb**							
	Baseline				Post USGB				Baseline				Post USGB			
	Digit	NRS	Colour	SpO_2	Digit	NRS	Colour	SpO_2	Digit	NRS	Colour	SpO_2	Digit	NRS	Colour	SpO_2
09/01/21	1	0	Tip black	100%	1	0	Tip black	100%	1	0	Tip black	100%	No block required			
	2,4,5	0	Normal	100%	2,4,5	0	Normal	100%	2,3,4,5	0	Normal	100%				
	3	3	Blue	100%	3	1	Blue	100%								
11/01/21	1	0	Tip black	100%	1	0	Tip black	100%								
	2,4,5	0	Normal	100%	2,4,5	0	Norma	100%								
	3	3	Blue	100%	3	0	Blue	100%								
	Right lower limb								**Left lower limb**							
	Baseline				Post Block				Baseline				Post Block			
	Digit	NRS	Colour	SpO_2	Digit	NRS	Colour	SpO_2	Digit	NRS	Colour	SpO_2	Digit	NRS	Colour	SpO_2
09/01/21	1,2,4,5	0-1	Black	NR	No block required				1,2,5	0	Black	NR	No block required			
11/01/21	3	0-1	Blue	97%					3,4	0	Blue	97%				
14/01/21	Bilateral foot debridement under SAB															

NR: Not recordable; IW: Irregular waveform; SpO_2:Oxygen saturation; USGB: Ultrasound-guided stellate ganglion block; NRS: Numerical rating scale ; SAB: Subarachnoid block.*: NRS/hour at which noted

reported in adults due to the risk of bilateral phrenic and recurrent laryngeal nerve palsy and breathing difficulty.[2,3] To the best of our knowledge, no similar case is reported in children. For safety, in the present child, all SGBs were performed under US guidance[4,5] and each block was performed after an adequate time gap to note any complications and haemodynamic instability.

Bilateral lumbar sympathectomy and epidural were not given for the lower limbs, due to large volumes of required local anaesthetic (LA) along with SGB and risk of LA toxicity. Thus, the total dose of LA was calculated at all times and all blocks were undertaken with monitoring and resuscitation equipment. The additive effect of analgesia with clonidine and vasodilatation secondary to LA leads to significant pain relief and reversal of vasospasm.[2,6]

Sympathetic blocks are invaluable to reverse pain and vasospasm in vasculitis. On late presentation, irreversible changes lead to auto-amputation; however, sympathetic blocks can halt the progression of gangrene, as was seen in the present patient.[2]

To conclude, in children with newly diagnosed MCTD and RP, sequential SAB, and bilateral USGB can be undertaken with careful planning and monitoring for successful remission of pain and vasospasm.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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