Effective plexus anaesthesia in a patient with Ehlers-Danlos syndrome type III

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Editor,

Ehlers–Danlos syndrome type III (also called hypermobility type) is an autosomal dominant-inherited disease of the connective tissue with an incidence of 1:150 000 [1]. Although defects in the genes encoding for procollagen type III (COL3A1) or tenascin-X (TNXB) have been suggested as possible candidates, up to now the precise defect for this genetic disease remains unknown [2]. From an anaesthesiologist’s point of view, the most important features in patients with Ehlers–Danlos syndrome are vessel fragility, poor skin healing, excessive bleeding, spontaneous pneumothorax, joint dislocation including vertebral instability, valvular prolapse and – in type IV – spontaneous dissections and ruptures of major vessels. Thus, the vessel fragility and successive bleeding as well as scoliosis may be a relative contraindication against regional anaesthesia in favour of general anaesthesia, whereas cervical spine joint laxity, skin fragility, an increased risk of temporomandibular joint dislocation, laryngeal trauma and pneumothorax may push the pendulum back in the direction of regional anaesthesia. Furthermore, there have been studies showing that subcutaneous infiltration of lidocaine has a reduced time of action and that transcutaneous application of a eutectic mixture of lidocaine 2.5% and prilocaine 2.5% (EMLA, AstraZeneca, Södertälje, Sweden) did not produce sufficient analgesia [3]. Additionally, patients with benign joint hypermobility syndrome (incidence 1:10 000), a syndrome with clinical features overlapping with those of Ehlers–Danlos syndrome type III, and a possibly similar genetic background [2] report an increased ratio of local anaesthesia failure compared with controls [4]. In contrast to these observations, there have been case reports of successful epidural and spinal anaesthesia in patients with Ehlers–Danlos syndrome [5–7].

To our knowledge, there are no reports on peripheral nerve blocks in patients with Ehlers–Danlos syndrome. This would be interesting, especially in patients with Ehlers–Danlos syndrome type III, as, in these patients, the ‘possibility of a resistance to local anaesthetics’ has been stated [4]. Furthermore, these patients undergo repeated operations for their unstable joints, and it would be valuable to have knowledge about the possibility of peripheral nerve blockades in these patients.

Case report

A 56-year-old female patient was scheduled to undergo a repeated arthroplasty of her left carpometacarpal joint. She was known to have a mitral valve prolapse, had

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some bleeding from the puncture site necessitating a contact with all visible vessels was avoided, there was of the local anaesthetic around the plexus. Although 1 ml lidocaine 1% in order to check adequate spread of a minuscule air bubble to visualize the end of the catheter. Twenty millilitres of mepivacaine 2% was injected above the tip of the needle (Contiplex D; B. Braun, Melsungen, Germany). Finally, the correct positioning of the catheter was checked by a test injection of 1 ml lidocaine 1% in order to check adequate spread of the local anaesthetic around the plexus. Although contact with all visible vessels was avoided, there was some bleeding from the puncture site necessitating a local compression for a couple of minutes. Thereafter, no further bleeding or swelling was seen.

The brachial plexus was blocked completely within 15 min, and the patient underwent the operation without any additional sedation or analgesia. She received satisfactory postoperative pain therapy via the brachial plexus catheter for 36 h (bupivacaine 0.125% with an infusion rate of 6 ml h\(^{-1}\), visual analogue scale at all times <20%) without any additional rescue medication and went home pain-free on the second postoperative day.

**Discussion**

This is the first description of peripheral nerve blocks in a patient with Ehlers–Danlos syndrome. The case shows that peripheral regional anaesthesia is an option in these patients making the ‘possibility of resistance to local anaesthesia’ [4] in this patient group unlikely.

Ehlers–Danlos syndrome is a heterogeneous group of inherited diseases characterized by abnormal connective tissue. Because of the life-threatening fragility of the great vessels in type IV, this form has received considerable attention from anaesthesiologists [7,8]. In contrast, type III has received little interest, though a reduced efficacy of local anaesthetics could be demonstrated. The infiltration anaesthesia with lidocaine is shortened and, with a eutectic mixture of lidocaine and prilocaine, sufficient analgesia could not be reached. Furthermore, 58% of patients with benign joint hypermobility syndrome (which is regarded by some authorities as identical to Ehlers–Danlos type III [2,9]) reported experiencing an ineffective local anaesthesia in the past, whereas this was reported by only 21% of the matched controls [4]. Thus, at least with transcutaneous and subcutaneous infiltration of local anaesthetics, there is a reduced efficacy. This may be due to the altered texture of the cutaneous and subcutaneous connective tissue leading to a faster spread or systemic resorption or both of local anaesthetics. In contrast, case reports have documented a normal effectiveness during spinal and epidural anaesthesia [5,7]. To our knowledge, this is the first case report of successful plexus brachialis blocks of normal duration in a patient with Ehlers–Danlos syndrome type III. Possibly in these locations, the distribution and or resorption of local anaesthetics are unaltered through the abnormal connective tissue. Certainly, the nerves of patients with Ehlers–Danlos type III or benign joint hypermobility syndromes are not ‘resistant’ to local anaesthetics as stated by some authors [4]. All voltage-gated sodium channels of eukaryotes have a highly conserved region that binds to local anaesthetics [10]. Beyond that, the genetic variations suggested to induce Ehlers–Danlos syndrome type III (mutations in procollagen III or in tenascin-X) have no influence on the sensitivity of the voltage-gated sodium channel. Thus, the reason for the decreased effectiveness of
transcutaneous or subcutaneous local anaesthesia is likely to be an altered pharmacokinetics rather than a change in pharmacodynamics.

Another problem in patients with Ehlers–Danlos syndrome is the fragility of blood vessels and the pleurae. Nowadays, injury to sensitive surrounding structures such as great vessels or the pleura may be avoided by the use of ultrasound-guided regional anaesthesia [11]. Naturally, damage to small vessels cannot be avoided and, therefore, our patient had a minor bleed next to the catheter at the end of the procedure. We chose an ultrasound-guided supraclavicular in-plane approach, because supraclavicularly the plexus brachialis is most superficial to the skin and the cannula can be visualized the whole way with minimal tissue damage. Furthermore, the neural structures are very compact there, avoiding multiple redirection of the needle as, for example, with an axillary approach. However, with an ultrasound-guided technique, the chance of injuring sensitive structures can only be lower but may never be avoided completely.

In conclusion, peripheral nerve blocks in a patient with Ehlers–Danlos syndrome type III worked as effectively as in normal persons, whereas infiltration anaesthesia – as described in the literature – is less effective. Ultrasound-guided regional anaesthesia may help to avoid injury to vessels or the pleurae, which are extremely fragile structures in these patients.

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