Letters

THE OCCULT SUBMUCOUS CLEFT – IMPROVING DETECTION BY EDUCATION

Editor,

Cleft palate is a congenital abnormality usually detected during the neonatal assessment. The most minor variant of cleft palate is known as a submucous cleft. Although submucous clefts share the feature of abnormal palate musculature, the overlying mucosa is intact and therefore they do not have the obvious gap in the palate associated with a complete cleft palate, which enables early diagnosis in the neonatal period. Calnan's triad of clinical signs are said to be diagnostic of a submucous cleft; Bifid uvula, zona pellucida and a notch in the hard palate, but these are not always present in all patients. (Figures 1 and 2).

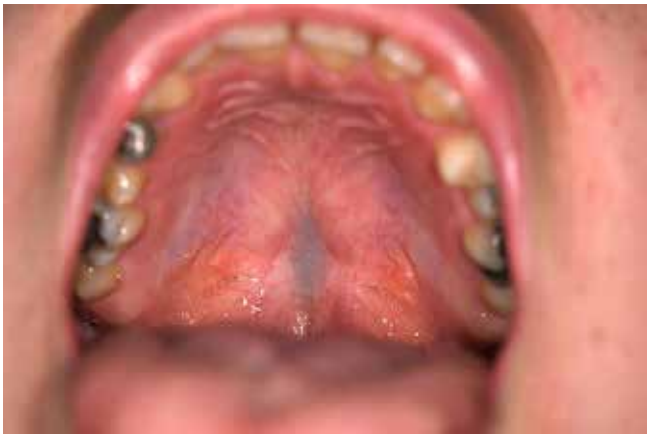

Fig 1. The zona pellucida – A blueish coloured area in the midline of the palate which represents the diastasis of the levator veli palatini muscles

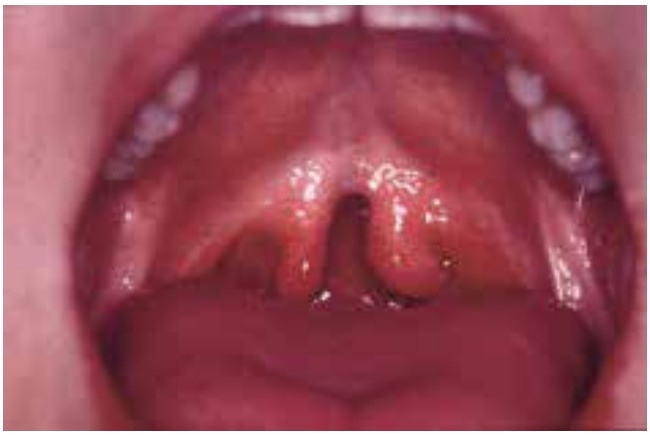

Fig 2. A bifid uvula

Children can have an occult submucous cleft with less than three of these features and some have none at all. Clinical diagnosis is understandably difficult and submucous clefts are detected at a much later stage when children are overtly symptomatic with speech and language difficulties around school age.

Symptoms can vary depending on the age of the child. In babies, feeding difficulties and/or nasal regurgitation are common. As the child gets older they develop recurrent ear problems, including recurrent otitis media with effusion and hearing problems. Speech and language problems become more apparent as the child develops and are caused by the abnormal positioning and insertion of the palate muscles. All of these symptoms present to the general practitioner in the first instance. This tends to be over multiple attendances during early childhood if these symptoms are problematic. This therefore provides a key opportunity for earlier recognition and diagnosis and therefore earlier treatment.

The importance of detection of children with a submucous cleft is to ensure appropriate intervention at an early stage prior to potentially irreversible speech and language problems. Not all patients are symptomatic and those with only mild symptoms may not require surgery. Speech and language therapy may be sufficient to normalise speech in a proportion of these children but this still requires recognition and referral to the cleft specialist speech therapists. For those that do require surgical intervention, prompt diagnosis and operative intervention will ensure speech outcomes can be optimised.

A recent paper by Baek *et al* has shown that speech outcomes following surgical intervention are better before the age of 5.5-years, highlighting the need for early diagnosis and treatment.[1]

We performed a retrospective review of children born with a submucous cleft in Northern Ireland. We found a significant increase in the number of patients with a submucous cleft over a 15-year period, from only 6 patients between 1988-1995 to 25 patients between 2003-2010. The average age for primary repair of the palate in the earlier cohort was 6 years which reduced to 5.2 years in the more recent cohort.[2] Highlighting once again that children with a submucous cleft in Northern Ireland are still being diagnosed at a late stage, when speech and language issues are hindering their progress during the early school years.

We urge a high index of suspicion for all general practitioners, paediatric and ENT specialists who are treating pre-school aged children with these symptoms. Any child presenting with repeated episodes of otitis media, nasal regurgitation or speech difficulties should prompt consideration of a diagnosis of submucous cleft. Examination of the palate may reveal the features described above and this warrants onward tertiary referral to the regional cleft team for further investigation and management.

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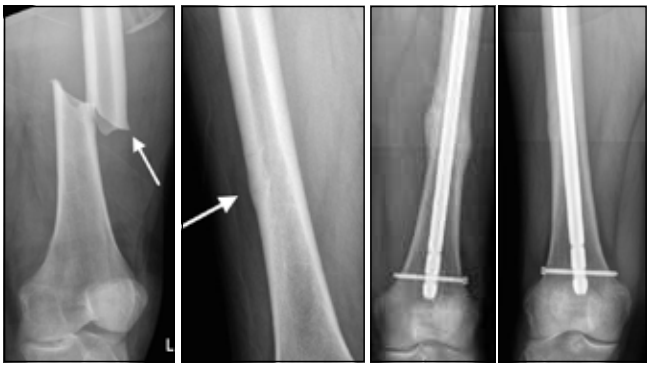
AN 'ATYPICAL' ATYPICAL FEMORAL FRACTURE

Editor,

A 75 year-old-male was admitted after sustaining a left femoral shaft fracture. He reported severe pain in his left thigh whilst descending steps followed by a sudden 'giving way' causing him to fall. Closer questioning revealed that he had been experiencing pain in his left thigh for approximately 8 weeks prior to the event but had no pain in the right thigh. His medical history included hypertension, hypothyroidism, reflux oesophagitis, type II diabetes mellitus and hypercholesterolaemia. Long-term medications included omeprazole, metformin and levothyroxine.

Left femur radiographs demonstrated a fracture at the junction of the middle and distal thirds with a short oblique pattern and localised periosteal thickening of the lateral cortex in keeping with an atypical femoral fracture [AFF] (**Figure 1a**). Admission blood and urine tests, including tumour markers, were unremarkable. A chest, abdomen and pelvic CT scan did not reveal any abnormality. Radiographs of the right femur demonstrated an incomplete fracture at the junction of the middle and distal thirds with focal thickening of the lateral cortex (**Figure 1b**). Antegrade intramedullary nailing was performed on the fractured side followed by prophylactic nailing on the contralateral side one week later. Metabolic bone assessment revealed a normal serum level of calcium, phosphate, parathormone and bone alkaline phosphatase with a slightly reduced vitamin D level, raised resorptive bone markers (C-terminal telopeptide, CTX) and low-level bone formation markers (N-terminal propeptide of type-I procollagen, P1NP). Bone densitometry was normal. Radiological fracture union was evident on both sides after approximately 10months (**Figure 1c and d**).



(a) (b) (c) (d)

Fig 1. (a) Anteroposterior radiograph (AP) of left femur- note short oblique fracture pattern with 'beaking' of the lateral cortex (white arrow); (b) AP radiograph of right femur demonstrating incomplete fracture with thickening of the lateral cortex (white arrow); (c) AP radiograph of left femur demonstrating complete union; (d) AP radiograph of right femur demonstrating complete union.

AFF's are defined as atraumatic or low-trauma fractures located between the subtrochanteric and supracondylar regions of the femur and have characteristic clinical and radiological features.[1] The American Society for Bone and Mineral Research (ASBMR) have proposed a set of specific criteria in order to identify AFF's with the requisition that at least four 'major criteria' should be observed.[2] In this case, there was no trauma, the fracture line originated at the lateral cortex with a transverse orientation, no comminution was present and there was localised periosteal thickening of the lateral cortex at the fracture site thus fulfilling the diagnostic criteria for an AFF.

The exact pathogenesis of AFF's is unknown. Bisphosphonate use is a key risk factor for AFF occurrence. [2,3] Other risk factors include genu varum, femoral bowing, collagen diseases and bone disorders characterized by low bone turnover, such as hypophosphatasia or pycnodysostosis.[4] Kim et al.[5] identified increased use of a proton-pump inhibitor (PPI) in AFF patients. The contralateral femur is affected in approximately 28 % of cases[2] and radiographic assessment is recommended even in the absence of symptoms. Intramedullary nailing is the treatment of choice for both complete and incomplete AFF's.[4]

This case highlights firstly, that AFF's can occur in the absence of anti-resorptive bone therapy, femoral malalignment or disorders of low bone turnover and secondly, the importance of assessment of the contralateral femur. Long-term use of a PPI may have been a contributory factor to the AFF's however mechanisms remain undetermined.

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PREVENTION OF PRESSURE ULCERS IN NASAL BRIDGE DURING NON-INVASIVE MECHANICAL VENTILATION. DISCUSSION OF RESULTS

Editor,

We would like to discuss the results achieved in the study by Bishopp et al. recently published in Ulster Medical Journal in which strategies for skin care during non-invasive mechanical ventilation were suggested.[1] The discussion of this study refers to a limited protective benefit of hyperoxygenated fatty acids in a study carried out by our team and also suggested that our sample was too limited to draw conclusions.[2]

We would like to refute the suggestion that our study, "Preventing facial pressure in patients under non-invasive mechanical ventilation: a randomized control trial" has a small sample size.[2] Our study had a methodology with calculation of statistical sample size [3], declared in the article and in previous protocols, by a previous piloting with 40 patients (10 in each group). The piloting allowed a pre-analysis for the size calculation of the effect estimated in 15.8%.

In the case of our clinical trial, the result of the number of the sample calculated is 152 patients in total among the four study groups, making replacement of the losses as can be seen in Fig. 1 of our article. The sample has been calculated so that the results can be considered, assuming the size of the effect described, a statistical power of 80% ($\beta = 0.20$) and a confidence level of 95% ($\alpha = 0.05$).

For all these reasons, we consider it important to emphasise our results and suggest care strategies based on the application of hyperoxygenated fatty acids and /or essential oils every 4-6 hours in the contact areas of the interface during non-invasive mechanical ventilation, following advice in recent results published on the same line. [2,4]

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PREVENTION OF PRESSURE ULCERS IN NASAL BRIDGE DURING NON-INVASIVE MECHANICAL VENTILATION. DISCUSSION OF RESULTS. AUTHORS' RESPONSE TO PEÑA-OTERO ET AL.

Editor,

We would like to thank Peña-Otero et al. for their attention to our paper on the preventative effect of hydrocolloid dressings on nasal bridge pressure ulceration in acute NIV in the UMJ[1]. It was not our intention to demean by any means the study by Otero et al.[2] by mentioning that it was a small sample study. We have used the expression 'small sample sizes' generically referring to two other papers[3,4] without any indication of the power analysis involved alongside the paper by Otero et al.[2] However, we would like to take this opportunity to clarify our standpoint on the question of sample size calculation in Otero et al[2]. A total sample size of 152 is determined to detect an effect size of 15.8% for the stated size and power. But this sample size is valid for comparing 76 subjects in 2 groups to be able to draw the conclusion that hyperoxygenated fatty acids (HOFA) is responsible for the preventative effect rather than split over 4 groups. The pairwise comparisons as stated in Otero et al.[2] require a larger sample size in each group to achieve the required power of 0.8. However, we strongly feel that the study in Otero et al.[2] is a significant study in the area of prevention of nasal bridge pressure ulceration and we are indebted to them for a pioneering research in the field of the practical application of NIV.

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