

Asymptomatic brown patches on the trunk

Question

A 6-year-old otherwise healthy boy presented with a one year history of asymptomatic brown patches on the trunk. The lesions were progressively spreading despite daily cleaning of

the skin with soap and hot water. The eruption had been treated with antifungal cream for one month, with no improvement. Physical examination revealed brown-reticulated scaly macular plaques on the abdomen (Fig. 1). There were no other skin lesions. What is the most likely diagnosis? (Answer on page 206)



Fig 1 Brown reticulated scaly macular plaques on the abdomen.

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Boy with cerebral palsy and severe malnutrition: Do not miss the mealtime!

Question

A 14-year-old boy with cerebral palsy, classified as Gross Motor Function Classification System 5,¹ was referred to our attention for poor weight growth during the last 3 years and compromised nutritional status: weight for age < -2.5 SD according to the

Gross Motor Function Classification System level-specific growth charts²; triceps skinfold < 5th centile for age and sex; and failure to thrive. Laboratory tests were normal except for a mild microcytic anaemia. Parents denied signs of gastroesophageal reflux disease or dysphagia; history of aspiration pneumonia was absent at anamnestic evaluation. A direct observation of mealtime by

physicians, however, highlighted cough, facial expressions of pain and significant drooling during feeding; a barium swallow test was therefore performed (Fig. 1). What is the diagnosis and how should this case be managed? (Answer on page 207)

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Fig 1 Barium swallow test showing immediate and bilateral barium inhalation and large hiatus hernia.

Scans propagating exponential workload (SPEW)

Answer

Hyperparathyroidism due to a parathyroid adenoma was the aetiology of this child's hypercalcaemia. The presence of a second lesion raised several possibilities, although given the proximity to the vasculature, consideration was given to paraganglioma, a group of neuroendocrine tumours that originate from either the parasympathetic or sympathetic nervous system. Paraganglial head and neck tumours are typically non-functional in children. If functional, mutations in the succinate dehydrogenase subunit account for most head and neck tumours. Other genetic syndromes in order of likelihood include MEN type 2, MEN type 1, von Hippel–Lindau syndrome and neurofibromatosis type 1. In this case, genetic testing for MEN2A, undertaken in the context of coexisting hyperparathyroidism, returned with no mutations detected.

Several imaging modalities are available and provide useful additional information for head and neck tumours, if interpreted within the clinical context. In this case, the ultrasonography characteristics, as well as findings of a T2-hyperintense contrast-enhancing lesion on MRI supported an initial diagnosis of

paraganglioma. However, literature has shown that over 90% of paraganglioma cases have positive DOTATATE uptake which was not observed in this case.¹

Review of the ultrasonography images at a multidisciplinary team meeting raised the question of whether the second lesion could be thymus. At the time of the left superior parathyroidectomy, a biopsy of the carotid sheath mass was pursued, with careful medical planning to confirm that the carotid sheath mass was non-secretory and surgical planning to avoid damage to surrounding structures. Histological analysis for the carotid sheath mass revealed thymic tissue.

The thymus is a lymphatic organ that arises from the third and fourth pharyngeal pouch, similar to the parathyroid glands. It migrates caudally along the thymopharyngeal duct, and ectopic thymic tissue can occur at various points along this descent, as well as around the major vessels.² Remnant cervical thymus can present at any age and has been reported in more than two-thirds of children; it tends to increase in size during childhood before involution occurs eventually in adolescence.^{2,3} Thymic remnants can form cysts during physiological atrophy, which

2 Vakirlis E, Theodosiou G, Lallas A, Apalla Z, Sotiriou E. Terra firma-forme dermatosis: Differential diagnosis and response to salicylic acid therapy. *Pediatr. Dermatol.* 2019; **36**: pde.13807.

3 Sechi A, Patrizi A, Savoia F, Leuzzi M, Guglielmo A, Neri I. Terra firma-forme dermatosis: A systematic review. *Int. J. Dermatol.* 2020; ijd.15301.

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Answer

A barium swallow test showed immediate and massive bilateral barium inhalation and a large hiatus hernia. A percutaneous endoscopy gastrostomy and laparoscopic Nissen fundoplication were performed with subsequent gradual improvement of clinical general condition, anthropometric parameters and quality of life of the child and his family. Prevalence of undernutrition in patients with cerebral palsy is still high world-wide and increases with more severe neurologic impairment, mainly due to feeding problem.³ Oropharyngeal dysfunction is the most common gastrointestinal difficulty with a prevalence up to 90% especially in more compromised children.^{3,4} Remarkably, typical signs of swallowing disorders such as long time-taking meals (more than 30 min), drooling, coughing, gagging and choking are sometimes underestimated by parents. This case highlight the importance of closed monitoring of these children with early recognition of malnutrition; direct observation of meal times is essential for a prompt diagnosis of dysphagia, even in the absence of characteristic signs at anamnestic evaluation, allowing specific treatment with clinical and life quality improvement in most cases.^{5,6}

Patient consent was obtained.

References

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- 6 Sullivan PB, Juszcak E, Bachlet AM *et al.* Impact of gastrostomy tube feeding on the quality of life of carers of children with cerebral palsy. *Dev. Med. Child Neurol.* 2004; **46**: 796–800.