



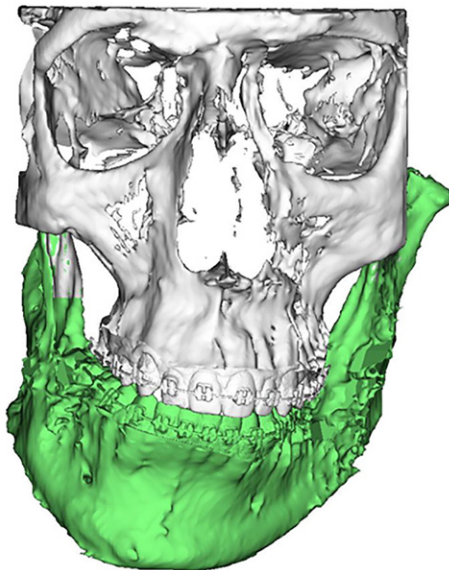
## Correspondence

**Neurofibromatosis type 1 and right mandibular hypoplasia: unusual diagnosis of occlusion of the left common carotid artery**


Dear Editor,

Neurofibromatosis type 1 also known as Von Recklinghausen's disease is an autosomal dominant neurocutaneous disease, with a disturbance of the NF1 gene localized in the 17th chromosome. Its incidence is 1 of 2500–3300 newborns. It is characterized by the formation of tumours in ectoderm and mesoderm tissues; consequently, its clinical manifestations are widely varied. Most frequent clinical manifestations are café-au-lait spots, axillary and groin freckling, Lisch nodules and neurofibromas. Other manifestations include hyperpigmentation, tumours and skeletal, neurological and cardiovascular disturbances [1]. Given the low incidence of this condition, and the lack of evidence-based systematized protocols calls for a meticulous anaesthetic evaluation and individualized planning [2].

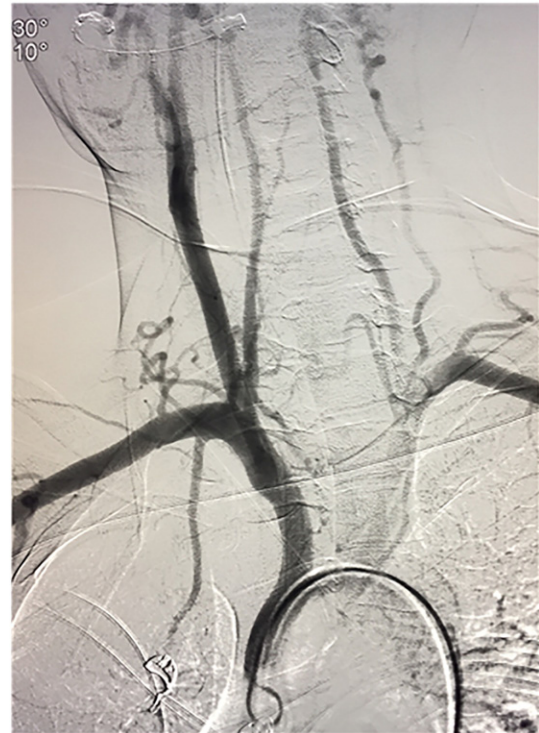
A case of a 18 years old teenager with neurofibromatosis type 1 (café-au-lait spots exclusively diagnosed) and right hemifacial hypoplasia (Fig. 1) is presented. He was scheduled for bimaxillary orthognathic surgery to correct his phacial dysarmony. Preoperative evaluation did not anticipate difficult airway management. Nevertheless, intravenous induction of anaesthesia and nasal intubation with videolaryngoscope (GlideScope - Verathon Medical Europe,



**Fig. 1.** Preoperative cone-beam computed tomography evaluation evidenced a right mandibular hypoplasia.

Amsterdam, Holland) was performed for direct vision of the airway in the context of a patient with neurofibromatosis 1 that can associate tumours of pharynx and larynx. After incision of yugal mucous of the mandibular vestibular cortical, a massive arterial bleeding was objectified. 1 L of blood was collected in the first 10 min, requiring invasive monitoring (Mostcare monitor-Pressure recording analytical method, PRAM; Vytech Health™, Padova, Italy), blood reserve and intubated transfer to the interventional neuroradiology unit.

Angiography study revealed an occlusion of the left common carotid artery from its origin (Fig. 2). Left brain hemisphere was being revascularized by intern carotid artery and extern carotid artery from anastomosis from the follow hypertrophied arteries (left ascendant cervical, left vertebral, left posterior communicant and anterior communicant complex). This situation, together with an important hypertrophy of the right extern carotid artery was partially balancing the contralateral occlusion. There were no vascular injuries like dissection or active bleeding points. A prophylactic supraseductive embolization in the intern maxillary artery (were the initial bleeding of the mandibular branch originated) was settled to avoid new bleedings and minimize ischemic risk.



**Fig. 2.** Anteroposterior angiographic study of the supra-aortic trunks revealed an occlusion of the left common carotid artery from its origin, with a hypertrophied left ascendant cervical artery.

The procedure proceeded uneventfully. The patient remained hemodynamically stable without needing vasoactive drugs or blood transfusion. He was extubated awake and transferred to postoperative care unit without further complications.

Surgery planning was changed after aberrant vascular anatomy of head and neck was diagnosed. To decrease intra and postoperative bleeding, a lower invasive surgery with distraction osteogenesis was made.

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### References

- [1] Hirsch NP, Murphy A, Radcliffe JJ. Neurofibromatosis: clinical presentations and anaesthetic implications. *Br J Anaesth* 2001;86:555–64.
- [2] Delgado JM, de la Matta Martín M. Anaesthetic implications of von Recklinghausen's neurofibromatosis. *Paediatr Anaesth* 2002;12:374.

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