Rendu-Osler Weber Disease (Rowd)
Alain Gerbaulet, Jean-Jacques Mazeron

1 Introduction

Rendu-Osler-Weber disease (ROWD) is an autosomally dominant inherited disease which affects the connective tissue of blood vessels. The primary symptom is haemorrhage, from the digestive tract (stomach), skin or nasal mucosa (3).

The most common presentation is recurrent epistaxis which may be managed by coagulation (diathermy or laser), systemic administration of estrogens, arterial embolisation (external carotid branches), skin grafting (dermoplasty) and brachytherapy (7).

Taking into account the very severe problems caused to the patients, brachytherapy is indicated in case of severe epistaxis after failure of other treatment modalities. Maylin has proposed a symptomatic classification for epistaxis (2).

2 Anatomical Topography

The internal part of the nose consists of two nasal fossae separated by a cartilaginous septum behind and under the bony structures of the nose. Each nasal fossa has a triangular form and is separated in four parts by the conchae. The lower part of the nasal fossae is called the floor, the upper part is delimited by the dorsum of the nose.

3 Pathology

The site of mucosal hemangiomas of ROWD is essentially along the very vascular mucosal lining of the nasal septum and thus constitutes the target volume.

4 Work Up

The symptoms of ROWD may appear at puberty and worsen episodically throughout life. Haemorrhagic telangiectasia, characteristic of this disease, can affect different integuments and mucosae (8).

Investigation must therefore include all areas which may be affected: skin (dermatologic exam), digestive mucosa (endoscopy), oral cavity and, of course, nasal fossae.

The major risk is haemorrhage and work-up investigation must include systematic blood tests and genetic family research.

5 Indications, Contra-indications

Brachytherapy is not indicated as a prophylactic treatment when the patient is asymptomatic (7).
Different therapeutic approaches may be tried in the management of epistaxis. Unfortunately, few are effective (4,5).

For recurrent severe epistaxis, aggressive treatments are justified, which include skin grafting, arterial embolisation and brachytherapy (2,7).

6 Target Volume

The target volume must encompass all the mucosa lining the nasal septum to a depth of 2 to 4mm. In case of bilateral disease, each nasal fossa must be considered and treated separately, not simultaneously. The most symptomatic side should be treated first. If treatment is needed to the other side, the contribution of irradiation delivered during the first therapeutic step must be evaluated (6,7).

7 Technique

For the brachytherapy implant, the plastic-tube technique is indicated. The patient is in a sitting position and an X-ray is taken in order to check the position of the tubes. Local anaesthesia is used (for example with xylocaine spray) and the tubes are positioned inside the nasal fossa: the lower tube close to the floor, the upper one following the antero-superior part of the nasal fossa and the last tube, between the other two. We recommend introducing the tubes in this order, as they should have a fan shape to cover the entire surface of the nasal mucosa of the septum (Fig 35.1). Since the three tubes diverge, tubes n°1 and n°2 are generally loaded for practically all their lengths and tube n°3 on its distal part (Fig 35.1) to try and deliver a homogeneous dose. At the end of the brachytherapy procedure, the different tubes are fixed at the nasal orifice by metallic buttons and strips.(2,5,6,7)

Fig 35.1A: Plastic tubes inside the nasal fossa, fixed with tape to the nose.
Fig 35.1B: Radiograph of plastic tubes in the nasal fossa. The different iridium loading according to the position of each plastic tube.

8 Dosimetry

The dose is calculated in 2 - 3 planes as perpendicular as possible to the radioactive lines. Using the Paris system, the reference isodose is by definition 85% of the basal dose.
Brachytherapy for ROWD is not an interstitial brachytherapy but a plesiobrachytherapy. The reference isodose must encompass the target volume, taking into consideration the depth under the surface mucosa at which the irradiation needs be delivered; therefore, if the 85% reference isodose is not considered to be adequate, the 80 or 70% isodose must be chosen (Fig 35.2), (2,3,7). The dose is systematically reported on the 85% reference isodose, but also on the isodose on which the dose is delivered (for example 80%, 70% ...).

![Computerized dosimetry: two plans, one sagittal (A), parallel to the iridium wires; one perpendicular (B) to the iridium wire (reference plan).]

9 Dose, Dose Rate, Fractionation

This LDR brachytherapy is given in one or possibly better two sessions. The total dose varies from 20 to 30 Gy at a dose rate of 8 to 12 Gy per day. The dose delivered to the opposite nasal fossa is routinely calculated, in case further brachytherapy is indicated (2, 3,7).

10 Monitoring

During the application performed under local anaesthesia, the tolerance of the patient is quite good. But because of the trauma caused by the tubes to the mucosa of the nasal-septum, epistaxis may be provoked, sometimes necessitating postponement of the procedure (2, 7).

During the one/two days of irradiation, the position of the tubes must be checked twice a day. Local treatments during this period are not usually needed (6).

11 Results

Very few data have been published on the role of brachytherapy in the treatment of ROWD. It is therefore difficult to be sure of its effectiveness, in this rare but often recurrent and severe affliction.

The largest series (7) reporting the Henri-Mondor Hospital experience includes only 43 patients. The conclusion of the authors appears optimistic when they suggest that: “intrasnasal brachytherapy is a
useful modality in the management of epistaxis in Rendu Osler disease” in comparison with their own results showing that out of only 15 patients evaluable for analysis, 13 had presented with recurrent secondary epistaxis. Nevertheless, it looks interesting to focus on the delay for recurrences, which ranged from 6 to 178 months.

In the Gustave-Roussy experience (1) of about 35 patients we have observed 70% of cases without recurrence during the first year after treatment.

Another publication coming from Pizzi et al.(6) showed that 4 out of 9 patients had a complete remission in 4/9 patients.

12 References