Schwannoma in Hemophilic Patient: Surgical Considerations

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ABSTRACT

Introduction: Schwannoma is a benign, encapsulated tumor that arises from Schwann cells of myelinated nerves. In this report, we present a case of schwannoma in a hemophilic patient and its surgical considerations related to bleeding complications.

Case Presentation: We present a case of a 27-year-old male coming to our center with a chief complaint of a painless, non-progressive growing mass on the back of his head since a year prior to admission. Physical examination showed that the mass was mobile, firm, and had a well-defined margin beneath the scalp. His radiograph showed a soft tissue mass beneath the scalp. MRI demonstrated a dense-multilobulated mass without intracranial infiltration. The patient had hemophilia A from the age of 5. We performed marginal excision of the mass with regiments of factor VIII (FVIII) concentrates preoperatively, intraoperatively, and postoperatively. Pathology was consistent with schwannoma.

Conclusions: Schwannoma management in hemophilic patients needs several considerations. Perioperative planning plays a major part in the management of patients with tumors and hemophilia to prevent bleeding complications.

INTRODUCTION

Hemophilia A is a bleeding disorder, described by factor VIII (FVIII) deficiency which results in prolonged bleeding, after surgery. The administration of FVIII prophylaxis is warranted to prevent bleeding in joints and muscles to avoid invalidating hemarthrosis and joint deterioration especially in both severe and some moderate patients. Administering FVIII concentrate is needed to obtain target trough levels above 1%. In the perioperative setting, physiological FVIII levels are required for a longer period [1]. Performing surgery in a hemophilic patient tends to be a management challenge for the surgeon [2].

Schwannoma is one of the most common peripheral nerve sheath tumors, which also include neurofibromas, perineuriomas, granular cell tumors, and malignant peripheral nerve sheath tumors [3]. Schwannoma is a benign, encapsulated, slow-growing tumor that arises from Schwann cells of myelinated nerves. It is mostly reported in the parapharyngeal region followed by paranasal sinuses, nasal cavity, scalp, submandibular region, larynx, epiglottis, and oral cavity. Some 25% to 45% of schwannomas are located in the head, and these often present as diagnostic and management challenges. The current treatment of choice is surgical and is often successful [4].

Surgical treatment in the hemophilic patient may result in adverse outcomes after surgery, such as acute bleeding, delayed bleeding, transfusion of blood products, inhibitor development, and thrombosis. However, there was no consensus or standard guidelines in FVIII regiment in the surgical management of benign neoplasm definitive surgery. We have already conducted other studies about the interdisciplinary management of hemophilic patients as our guidelines [5]. The surgical management of schwannoma in hemophilic patients requires extensive surgical considerations and preparation considering the risk of complications. We present a 27-year-old male diagnosed with schwannoma in the occipital region and hemophilia A treated with surgical intervention. Written informed consent for patient information and images to be published was provided by the patient.

CASE PRESENTATION

A 27-year-old male was referred to our center with a mass on the right-back of his head a year prior to admission. The mass was slowly getting bigger until reaching the size of a tennis ball. There was neither tenderness nor wound or sinus from the lump (Figure 1). No other mass appeared elsewhere. The patient has
had Hemophilia A since the age of 5 years. The blood test showed a hemoglobin level of 12 g/dL, hematocrit of 39%, white blood cell count of 10,230/mL, platelet count of 545x10³/mL, prothrombin time of 10.2 seconds (control 10.7 seconds), aPTT 143.1 seconds (control 32.4) ESR 50mm, and FVIII level of < 1% with negative anti-FVIII (inhibitor).

The plain radiograph showed a soft tissue mass outside of the occipital bone without the bone cortex erosion (Figure 2). Magnetic resonance imaging (MRI) examination showed a well-defined soft tissue, a multilobulated mass beneath subcutaneous tissue, and no intracranial infiltration. Clinical and radiological results showed the benign nature of the soft tissue lesion raising the suspicion of schwannoma.

In the outpatient settings, this patient has already been given a maintenance dose of FVIII concentrates 1,000 IU once in two weeks. He underwent perioperative management of FVIII concentrates for surgical planning. He was administered with 2,250 IU FVIII concentrates 2 hours before surgery and 1,500 IU 12 hours postoperatively. From the first until the third postoperative day, the FVIII concentrates were given twice a day with a dose of 1,500 IU. In the fourth until the seventh postoperative day, the FVIII concentrates were given once a day with a dose of 1,500 IU. The patient was discharged on the eighth postoperative day. The management was continued in the outpatient setting with 1,500 IU FVIII concentrates three times a week.

The patient was in a prone position, and a straight incision was performed for marginal excision of the tumor. The mass was an encapsulated-multilobulated fat-like tumor, as shown in (Figure 3). No bleeding and infection complications were found. Histopathological examination confirmed the diagnosis of schwannoma (Figure 4).

Figure 1. The clinical picture of the mass. It was a no tender and mobile mass with size 9x8x4cm

Figure 2. A. Plain radiographs showed soft tissue mass outside of the occipital bone without the bone cortex erosion, B. MRI examination showed a well-defined soft tissue, multilobulated mass beneath subcutaneous tissue, and no intracranial infiltration.

Figure 3. Macroscopic view of the tumor. The mass was an encapsulated-multilobulated fat-like tumor

Figure 4. (A) Antoni A areas; (a). Verocay body of palisaded Schwann cells, (B) Antoni B area
DISCUSSION

In our case, we reported a 27-year-old with a predilection of tumor in the subcutaneous tissue of the occipital area of the head. He was also a hemophilia patient who could have complications such as bleeding and pseudotumor [1]. The patient’s percentage of coagulation factor was less than 1% and was categorized as severe hemophilia. Schwannoma can affect all ages, has no predisposition of sex, is mostly solitary, and mostly occurs in the peripheral nerve in the skin and subcutaneous tissue of the head and neck [6]. The MRI examination showed a well-defined soft tissue, a multilobulated mass beneath the subcutaneous area of the head with no cranial and intracranial infiltration. The MRI excluded the possibility of pseudotumor and malignancy. Clinical and radiological results showed the benign nature of the soft tissue lesion raising the suspicion of schwannoma. However, the final diagnosis was established after we performed surgical excision and clinicopathological conference with the radiology and histopathology department. We found that the mass was an encapsulated and multilobulated fat-like tumor. This result mirrors the literature in which, macroscopically, the tumor is solitary and globose, has a smooth surface and the size of less than 10 cm, and is encapsulated. Gross pathology of the tumor revealed firm, light tan glistening tissue, interrupted by white/yellow areas or patches of hemorrhage [3]. Microscopically, the mass consists of compact cellular lesions with interlacing and cellular fascicles (Antoni A) and fewer cellular and myxoid areas (Antoni B) [7]. Cells of Antoni A tissue possess modest amounts of eosinophilic cytoplasm, no discernible cell borders, and normochromatic elongated tapered nuclei. Cytoplasmic nuclear inclusions, nuclear pleomorphism, and mitotic figures may be seen. Palisading (verocay bodies) takes the form of parallel rows of Schwann cell nuclei separated by their aligned cell process. Antoni B tissue commonly contains a collection of lipid-laden histiocytes and thick-walled, hyalinized blood vessels [3].

Treatment of schwannoma is almost entirely surgical and is often successful except for possible damage to nerves as a result of its structure and to bones when it weakens the periosteal structure and causes fractures [4]. However, the high rate of local recurrence after inadequate excision has been previously emphasized. Radical local excision in a relatively early case gives the best chance of cure [8]. In our case, we performed marginal excision of the tumor with the diathermia technique.

There was a need for special consideration in surgery for the patient with hemophilia. Several decades ago, there were limited published data from developing countries, confirming the gross underdiagnosis of this condition in many parts of the world [2]. Operative management requires a multidisciplinary team with experience in the management of rare bleeding disorders. Our patient was treated with the FVIII concentrate protocol where we gave FVIII concentrates 2 hours before surgery, 12 hours after surgery, followed by maintenance dosing in the first 7 days postoperatively, and continued to the outpatient setting treatment three times a week [5].

There is no guideline for FVIII administration for surgical management in patients with hemophilia and tumor. However, previous studies have shown the benefits of FVIII concentrate administration on bone tumors and hemophilia. Two reports by Kamal et al. have demonstrated the practice of FVIII concentrate administration prior to the surgical treatment of pseudotumor [5,9]. In their study, FVIII concentrate was administered 1 hour before surgery to increase the level to 100% [9]. The post-surgical follow-up showed good clinical and functional outcomes up to 1 year after surgery [9]. Regiment throughout the perioperative and postoperative period is important, particularly in major surgery [10]. A previous study about preoperative administration of coagulation factors reported that coagulation factor administration should be adjusted with the condition of the patient. Bolus injection (BI) method or continuous injection (CI) method for hemophilia is recommended. The BI method uses bolus injection of coagulation factor when coagulation factor level reaches 50%. However, the fluctuation of the coagulation factor level and the risk of overdose could increase the risks of complications. The CI method is currently recommended for major surgery with a consistent level of the coagulation factor. However, the CI method usually requires specialized expertise which is not available [11].

Ideally, in preparing patients with hemophilia A for surgery, FVIII levels are routinely raised to approach 100% of normal activity. It should be maintained for the first 3 postoperative days. From the 4th day onwards, it should be maintained at 80%. From the 77th day onwards, it is allowed to decline to 40% of normal activity [12]. A previous study has reported the use of FVIII concentrates in a patient with Von Willebrand Disease (VWD) and hemophilia who underwent a surgical procedure and has shown sufficient results in maintaining hemostasis. A proposed dose of 35 IU/kg preoperatively and 10 to 20 IU/kg after that is sufficient for surgical hemostasis in this patient [2].

In this case, there were neither intraoperative nor postoperative complications. After 6 months of follow-up, there was no recurrence of tumor in the patient. He did not have further complaints or symptoms.
CONCLUSIONS

Perioperative planning plays a major part in the management of patients with soft tissue tumors including schwannoma and hemophilia to prevent bleeding complications. An appropriate diagnosis and complete surgical resection of this tumor provide bleeding prevention, symptoms relief, and esthetical improvement in the affected region.

DECLARATIONS

Competing of Interest

The authors declare no competing interest in this study.

Acknowledgment

Not applicable

REFERENCES