

Introduction

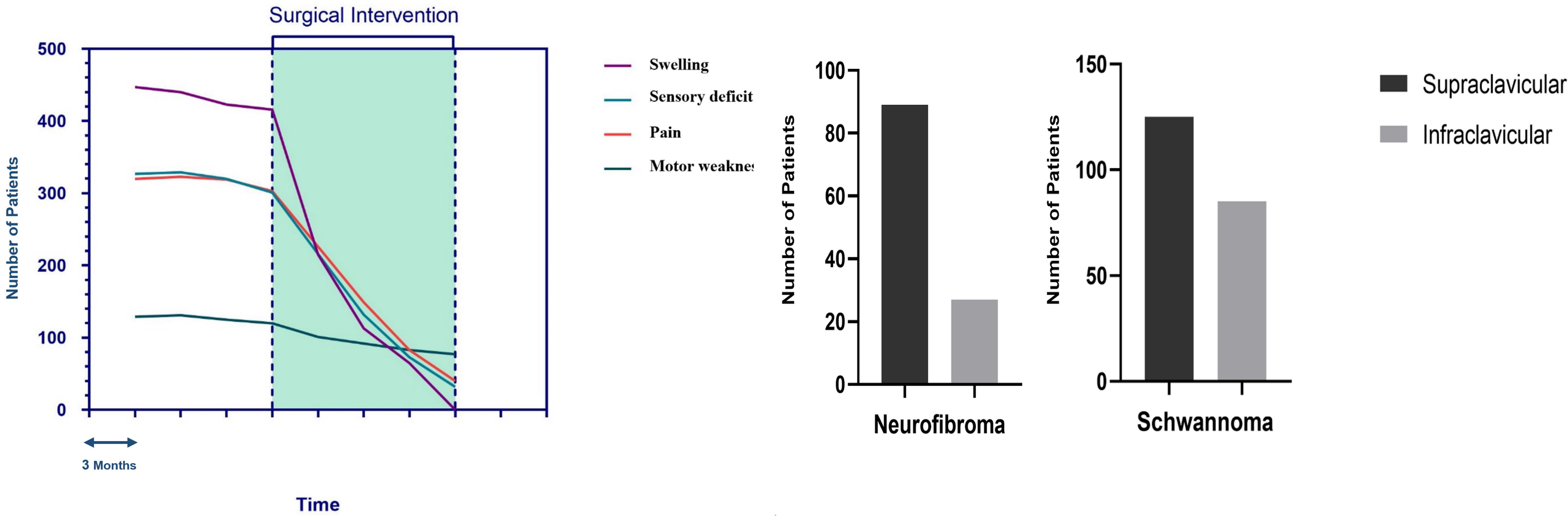
Primary brachial plexus (BP) tumors are rare, challenging-to-treat entities that mainly originate from the neuroectodermal cells. These constitute nearly 5% of all upper extremity tumors, causing pain and disability in affected patients. Clinical presentation of BP tumors can differ depending on their location and size. Most often they present with a painful palpable mass with or without neurologic deficits. This is the most common symptom of patients with BP tumors. As the tumor progresses, painful paresthesia at the distribution of the affected nerve occurs. Ultimately, progressive loss of motor function may occur. Various treatment options have been introduced for the management of primary tumors of the brachial plexus (BP), ranging from conservative therapy to wide local excision with/without postoperative chemoradiotherapy. However, there is a discrepancy about the optimal treatment strategies for benign and malignant lesions based on pathologic characteristics and the location of the BP tumors. Also, no consensus has been established to describe the optimal operative approach for tumors of the brachial plexus. Hence, this systematic review was performed to evaluate the optimal surgical interventions according to the pathologic characteristics of the tumor and describe overall patient outcomes, survival rates, and possible prognostic factors

Method

The literature search was conducted by the two reviewers using the four main online databases including Web of Science (WOS), PubMed, Scopus, and Google Scholar with the aim of assessing all related articles addressing the clinical outcome and role of surgical interventions for the management of primary tumors of the brachial plexus. All articles from 2000 until July 2022, were included in the study. The inclusion criteria for study selection were human studies of all age groups describing the clinical outcomes and optimal management of primary brachial plexus neoplasms (regardless of the tumor type) which were published in the English literature. Additionally, manuscripts containing non-primary neoplasms (metastatic), cadaveric, non-human studies, and studies without quantitative data (including case reports and technical notes) were excluded. The study findings were summarized descriptively and presented as means and standard deviation (SD).

Results

A total of 687 patients and 693 tumors were evaluated, in which the mean age of the participants was 41.7 ± 8.7 years old (range: 1-92 years). There were 298 (46.2%) males and 347 (53.8%) females. Of the 687 patients analysed, 79 (11.5%) patients had a history of neurofibromatosis and only 1(0.1%) patient had a history of schwannomatosis. The most common clinical symptoms were swelling (447, 65.1%), pain (320, 46.6%), sensory (327, 47.6%), and motor deficits (129, 18.8%). The mean duration of symptoms before medical intervention was 25.2 ± 12.3 months. Of 693 tumors, 629 (90.8%) tumors were benign and 64 (9.2%) were malignant, with a mean tumor size of 5.4 ± 3.1 cm. Of all tumors treated, benign tumors included 421 (60.8%) schwannomas, 126 (18.2%) neurofibromas (NF), 17 (2.5%) desmoid tumors, and 14 (2.0%) lipomas, as well as smaller numbers of ganglioneuroma, cystic hygroma, hemangioma, neurogenic cyst, and aneurysmal bone cyst. Malignant lesions included 48 (6.9%) malignant nerve sheath tumors (MNST), as well as 4 (0.6%) malignant granular cell tumors, 3 lymphomas (0.4%), 2 (0.3%) synovial sarcomas, and smaller numbers of the peripheral primitive neuroectodermal tumor, papillary carcinoma, and other sarcomas of the brachial plexus.



The trunks were the most common location for tumor involvement with 210 (37.9%) cases, followed by 202 (36.5%) involving the nerve roots, 124 (22.4%) tumors involving the cords, and 17 (3.1%) involving the terminal branches. The upper trunk (79, 55.6%) was the most common location followed by the middle (36, 25.4%) and lower trunks (27, 19.0%). For patients with nerve root involvement, most patients had tumors affecting the C7 nerve root (48, 31.0%), followed by the C5 (36, 23.2%) and C6 (33, 21.3%) roots. Gross total resection (GTR) was performed for the majority of the cases (432, 79.9%) and subtotal resection (STR) was performed in (109, 20.1%) patients. For patients with schwannomas (n=344), GTR was successfully achieved in 341 (99.1%) and only 3 patients (0.9%) underwent STR. In contrast, for patients with neurofibromas (n=109), GTR was performed for only 38 (34.9%) and 71 (65.1%) who underwent STR. Out of the 48 patients with the diagnosis of MPNST, 16 (50.0%) patients underwent extended surgical excision with wide margins and 16 (50.0%) underwent STR. For malignant tumors, a total of 14 (2.0%) patients underwent adjuvant radiotherapy following the surgery. The mean duration of follow-up was 31.1 months. Out of the 320 patients who presented initially with pain, 43 (13.4%) patients complained of remaining symptoms following the surgery. Also, 295 (90.2%) of patients with sensory deficits were symptom-free at the last follow-up. However, motor weakness remained for 79 (61.2%) of patients following the surgical interventions. Reviewing long-term outcomes (n=687), local tumor recurrence occurred in 15 (2.2%) patients, and distant metastasis was observed in only 8 (1.2%) cases. The overall mortality was 21 (3.1%) patients among the study population.

Conclusion

The degree of surgical excision (total or subtotal) for primary BP tumors mainly relies on the pathological characteristics, location, and extent of the tumor. Even with STR, patients with neurofibromas have excellent clinical outcomes with a very low rate of recurrence. In contrast, MPNSTs are highly aggressive tumors and are associated with an extremely poor outcome, even when complete resection is achieved