CONTENTS

Editorial
Criteria for deciding cost-effectiveness for expensive new anti-cancer agents
   Rajiv Sarin.............................................................................................................................................................1

Original Articles
The effect of three mouthwashes on radiation-induced oral mucositis in patients with head and neck malignancies: A randomized control trial
   PD Kumar Madan, PS Sequeira, Kamalaksha Shenoy, Jayaram Shetty .............................................................3
Implications of contrast-enhanced CT-based and MRI-based target volume delineations in radiotherapy treatment planning for brain tumors
   Niloy R Datta, Rajasekar David, Rakesh K Gupta, Punita Lal..............................................................................9
Radiofrequency ablation of hepatic metastasis: Results of treatment in forty patients
   GK Rath, PK Julka, S Thulkar, DN Sharma, Amit Bahl, S Bhatnagar.................................................................14
Execution of mantle field with multileaf collimator: A simple approach
   Ramachandran Prabhakar, Kunhi Parambath P Haresh, Pappiah S Sridhar, Macharla A Laviraj,
   Pramod K Julka, Goura K Rath...........................................................................................................................18
Prognostic and diagnostic value of serum pseudocholinesterase, serum aspartate transaminase, and serum alanine transaminase in malignancies treated by radiotherapy
   Arun Chougule, Sofia Hussain, Dwarka Prasad Agarwal .................................................................................21

Review Article
An overview on applications of optical spectroscopy in cervical cancers
   C Murali Krishna, GD Sockalingum, MS Vidyasagar, M Manfait, Donald J Fernandes, BM Vadhiraja,
   K Maheedhar.......................................................................................................................................................26

Case Reports
Radiotherapy for management of skin cancers in fibrodysplasia ossificans progressiva: A case report and review of the literature
   John Antony Frew, Charles G Kelly ....................................................................................................................37
Sarcomatoid squamous cell carcinoma of uterine cervix: Pathology, imaging, and treatment
   Milind Kumar, Amit Bahl, Daya Nand Sharma, Shipra Agarwal, Dhanapathi Halanaik, Rakesh Kumar,
   Goura Kishore Rath ............................................................................................................................................39

Brief Communications
Chest wall metastasis from hepatocellular carcinoma in the absence of a primary: An unusual presentation
   Kaustav Talapatra, Reena Engineer, Jai Prakash Agarwal, Shilpa Vyas, Shyam Kishore Shrivasvasta ..........42
Endobronchial metastasis of follicular thyroid carcinoma presenting as hemoptysis: A case report
   RAS Kushwaha, Sanjay Kumar Verma, Sanjay Vineet Mahajan........................................................................44
Accelerated partial breast irradiation: An advanced form of hypofractionation
   Ashwini Budrukkar ..............................................................................................................................................46
Coexistence of carcinoma breast and Paget’s disease of bone
   S Sundaraiya, PK Pradhan, A Gupta, M Jain, SK Mishra, BK Das.....................................................................48

Letter to Editor
Dysplastic hematopoiesis and underlying dysthyroidism
   Riad Akoum, Michel Saade, Wafic Tabbara, Emile Brihi, Marwan Masri, Khaled Habib, Gerard Abadjian......50

Reviewers’ List, 2007 .................................................................................................................................................51

The copies of the journal to members of the association are sent by ordinary post. The editorial board, association or publisher will not be responsible for non-receipt of copies. If any of the members wish to receive the copies by registered post or courier, kindly contact the journal’s / publisher’s office. If a copy returns due to incomplete, incorrect or changed address of a member on two consecutive occasions, the names of such members will be deleted from the mailing list of the journal. Providing complete, correct and up-to-date address is the responsibility of the members. Copies are sent to subscribers and members directly from the publisher’s address; it is illegal to acquire copies from any other source. If a copy is received for personal use as a member of the association/society, one cannot resale or give-away the copy for commercial or library use.
Case Report

Radiotherapy for management of skin cancers in fibrodysplasia ossificans progressiva: A case report and review of the literature

ABSTRACT

Fibrodysplasia ossificans progressiva (FOP) is a rare condition of ectopic calcification leading to increasing disability throughout life, with most patients being wheelchair bound by the age of 30. Ectopic calcification can be triggered by trauma and it is therefore important to minimize biopsies and operative procedures in affected individuals. We report a 46-year-old FOP patient who was successfully treated with radiotherapy for a basal cell carcinoma. There are no previous reports in the literature on the management of skin malignancies in these patients and very limited literature on outcome following external beam radiotherapy.

KEY WORDS: Basal cell carcinoma, fibrodysplasia ossificans progressiva, radiotherapy

INTRODUCTION

Fibrodysplasia ossificans progressiva (FOP), also known as myositis ossificans progressiva, is a rare condition of ectopic ossification, with only around 300 known patients worldwide. There are no reports on the management of skin malignancies in these patients and very limited literature on outcome following external beam radiotherapy.

CASE HISTORY

A 46-year-old gentleman with a background of severe FOP, presented in April 2006 with a basal cell carcinoma (BCC) involving the right upper lip. The diagnosis was made on clinical grounds, with no biopsy being performed because of the risk of heterotopic bone formation following trauma. He was treated with superficial x-ray radiotherapy (90 kV) to a dose of 35 Gray delivered in five fractions on consecutive days. The patient had a complete response to radiotherapy, with a good cosmetic result; there was no evidence of heterotopic bone formation [Figure 1].

DISCUSSION

FOP has autosomal dominant inheritance with complete penetrance but variable expressivity, and most cases result from sporadic mutation. Symptoms tend to begin in early childhood with tender swellings on the head, neck, or back. The diagnosis is supported in almost all patients by congenital malformations of the great toes. The malformations are recognizable at birth, and consist of a hypoplastic proximal phalanx with associated hallux valgus. Although ossification may not be evident early in the course of the disease, the swellings most often progress to ossification. The most common sites of early heterotopic ossification are the neck, spine, and shoulder girdle. Ossification tends to proceed in a cranial to caudal and proximal to distal pattern. The majority of patients have severely restricted movement of the upper limbs by the age of 15, with many confined to a wheelchair by 30 years of age.

The genetic mutation responsible for FOP has recently been mapped to chromosome 2q23-24 by linkage analysis in five families with unambiguous features of FOP. This revealed an identical heterozygous mutation (617→A; R206H) in the glycine-serine (GS) activation domain of ACVR1, a bone morphogenetic protein (BMP) type 1 receptor, in all the affected individuals examined. Protein modeling predicts destabilization of the GS domain, which is consistent with constitutive activation of the ACVR1 as the underlying cause of the ectopic chondrogenesis, osteogenesis, and joint fusions seen in FOP.

There is no effective treatment and it is, therefore, important to avoid soft tissue trauma, including biopsies, intramuscular injections, surgical...
procedures, and mandibular blocks for dental procedures; viral illnesses must also be avoided. All of these are recognized factors that may precipitate episodes of rapidly progressive heterotopic ossification, with resultant permanent loss of function in the affected area. Early diagnosis and avoidance of trauma are the key principles in the effective management of these patients. There are no reports of the use of liquid nitrogen cryotherapy for the treatment of skin cancer in FOP patients. However, in view of the potential for this modality to cause significant tissue inflammation there is always the fear that this could result in further heterotopic ossification.

A search of the PubMed database (of the National Library of Medicine) and Ovid MEDLINE did not locate any previous reports on the management of BCC in FOP patients. It is intuitive that the most appropriate management should be nonsurgical. Previous reports in the literature reveal similar outcomes following surgery or radiotherapy in the treatment of BCC.

Kitterman et al. sent a questionnaire to all 269 patient-members of the International FOP Association which is believed to include over 90% of all known FOP patients worldwide. Of the 138 responders, 87% of patients were initially given incorrect diagnoses, the commonest of which was cancer (32%). Of these patients, seven received radiotherapy but the authors do not comment on the outcome.

There are two other case reports in the literature of treatment with radiotherapy in patients with FOP. Radiotherapy was given to prevent ossification following the excision of heterotopic bone from the ankylosed hip joint of an 18-year-old FOP patient. There was a small amount of ossification at the operative site at 1 year, with a report of a good functional outcome. Radiotherapy was also effective in reducing the neuropraxia associated with an iliopsoas mass compressing the femoral nerve in an FOP patient.

It is the opinion of the authors that fractionated radiotherapy should be considered the standard of care for FOP patients with BCC and that biopsy should be reserved for those patients in whom the clinical diagnosis is uncertain.

REFERENCES


Source of Support: Nil, Conflict of Interest: None declared.