Acinic Cell Carcinoma of parotid gland with locoregional metastasis in young male

Authors: Dr. Anuradha Kusum*, Dr. Sushil Kumar Shukla2, Dr. Nadia Shirazi3, Dr. Dushyant Singh Gaur4, Dr. S.S. Bisht5,

1Professor and Head, Department of Pathology, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Dehradun, Uttrakhand (India).
2Senior Resident, Department of Pathology. Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Dehradun, Uttrakhand (India)
3,4Professor, Department of Pathology, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Dehradun, Uttrakhand(India)
5Professor and Head, Department of ENT, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Dehradun, Uttrakhand(India)

*Corresponding Author: Dr. Anuradha Kusum*

E-mail: anuradhakusum@srhu.edu.in

Abstract:
The Acinic cell carcinoma(ACC) is slow growing, low grade epithelial neoplasm of salivary glands, most frequently occur in parotid gland and preferentially more in female with median age at time of presentation is 52 years. FNAC may be useful tool for confirmation of the diagnosis. The treatment of tumor is essentially by surgical excision of gland. The most common his to-pathological presentation of tumor is solid sheet and nesting pattern of growth. The present case of 18 years old male which initially present as benign cystic lesion of parotid gland and the final diagnosis was ACC which metastasized to loco-regional lymph nodes. To conclude, ACC is frequently arising in parotid gland and metastasis to the intra-salivary gland lymph nodes is very rare finding in young male. Surgery with adjuvant post-operative radiation therapy is acceptable for final outcome of disease and the tumor should be managed uncompromisingly.

Keywords: ACC: Acinic cell carcinoma, Loco regional metastasis, Parotid gland
1. Introduction:
Acinic Cell Carcinoma (ACC) Is A Slow Growing And Low Grade Epithelial Neoplasm Of The Salivary Glands, Having Low Malignant Potential, In Which Few Of The Tumor Cells Exhibiting Acinar Cell Differentiation (1). ACC Was Firstily Identified As Pathological Entity by Godwin Et Al In Year 1954 (2). Most Frequently ACC Occur In Parotid Gland And Preferentially More In Female With Female: Male Ratio Is 3:2 And Median Age At Time Of Presentation Is 52 Years With Slightly More Than 16% Of Cases Were Under The Age Of 30 Years (3). The Loco-Regional And Distant Metastasis, Higher Tumor Grade And Larger Size Of Growth, Were More Commonly Seen Among Patient’s Age More Than 30 Years, Are Associated With Tumor Recurrence And Poor Prognosis(4). Fine Needle Aspiration Cytology (FNAC) Examination May Be Useful Tool For Confirmation Of The Diagnosis. The Treatment Of Tumor Is Essentially By Surgical Excision Of The Gland (5). In Histo-Pathological Examination; the Most Common Presentation of Tumor Is Solid Sheet And Nesting Pattern Of The Growth Followed By Microcystic, Papillary-Cystic And Follicular Pattern (6). The Purpose Of The Present Case Is To Present The Clinical, Cytological And Histopathological Features Of Of Acinic Cell Carcinoma Of The Parotid Glands Of 18 Years Old Male Which Initially Present As A Benign Cystic Lesion Of Parotid Gland Than Finally Diagnosed As ACC With Metastasized To Loco-Regional Lymph Nodes.

2. Case Report:
A 18 Years Old Male Patient Was Apparently Asymptomatic 5 Months Back When He Developed Swelling In Front Of The Right Ear Which Was Gradual In Onset, Progressively Increasing In Size. There Was No History Of Fever, Difficulty In Swallowing Or Breathing, Ear Discharge, Nasal Discharge Or Decreased Hearing. On Local Examination: Approximately 4x3 Cm Palpable Lump Below Right Ear Extending Upto Posterior Mastoid Tip. The Lump Was Cystic In Consistency, Non Tender With Overlying Normal Temperature With No Significant Palpable Cervical Lymphadenopathy. On USG Neck Finding Suggestive Of Multicystic Areas With Moving Internal Echoes Within Right Parotid Gland With A Necrotic Level I B Lymph Node Likely Infective ?Tubercular Etiology. MRI Face Showed Enlarged Right Parotid Gland With Multiple Cystic Lesions, Right Level IB And Bilateral Level II Cervical Lymphadenopathy. On FNAC, 17ml Fluid Was Collected And Cytological Smears Show Cells Are Arranged In Clusters And Papilliform Structure. The Tumor Cells Are Large With Mildly Pleiomorphic Nucleus, Few Of Them Are Eccentric, Showing Moderate Anisonucleosis, Course Chromatin And Small Nucleolus With Abundant Cytoplasm, Showing Vacuolations And Pink Granularity. Few Multinucleate Giant Cells With Abundant Pink Amorphous To Granular Material And Abundant Macrophages Are Also Seen. FNAC Finding Shows Features Of Neoplasm with Differential Diagnosis Of ACC And Mucoepidermoid Carcinoma. Patient Underwent For Right Parotidectomy Under GA. On Gross Examination, Parotid Gland Measures 5.5x4.0x0.5cm With Bosselated Outer Surface. Cut Surface Shows Multiple Cystic Cavities, Largest Measuring 1.2x1.0x0.2 Cm. On Sectioning Of Attached Fat, 4 Lymph Nodes Are Identified. On Microscopic Examination, A Multicentric Poorly Encapsulated Carcinoma Forming A Microcystic Pattern. Cells Have Round To Pleomorphic Nuclei With Variable Amount Of Clear Cytoplasm. Many Cells Show Intracytoplasmic Clearing And Mucin Deposition. Lymphovascular Emboli Are Present. Four Out Of Total 10 Lymph Nodes Shows Metastatic Deposits Of Carcinoma With Perinodal Extension. On Special Stain, PAS And PAS-D Stain Shows Focal Intraluminal And Intracytoplasmic Mucin Positivity. Final Impression Was ACC-Microcystic Type With Regional Lymph Node Metastasis. On Follow Up Patient Advised For Adjuvant Radiotherapy.
Fig 1: MGG Stain on Cytological Smears Showing Malignant Tumor (400x)

Fig 2: HE Stain on Histological Section of Lymph Node Shows Metastatic Deposits of Acinic Cell Carcinoma with Perinodal Extension (200x)

3. Discussion:
ACC of Salivary Gland Was First Described In Literature by Nasse In Year 1892 As A Benign Tumor And As Clinic-Pathological Entity By Godwin In Year 1954 (2). Most Frequently, ACC Occurs In The Parotid Gland With Other Unusual Primary Sites Were Oral Cavity, Lips, Hard Palate, Larynx, Mandibular Bone, Nasal Cavity, Paranasal Sinuses, Pancreas, Stomach, Lung, Breast And Prostate (7). The Hazardous Factors That Can Causes ACC Include Radiation Exposure; Industrial Workers Include Rubber And Asbestos Industries, Plumbing Industries, And Automobile Industries. The Etiopathogenesis Shows Strong Relationship between Epstein Barr Virus (EBV) and Lympho-Epithelial Carcinoma (3). Endogenous Hormone Receptors Of Androgen, Estrogen And Progesterone Were Reported In Previous Literature In A Small Number Of Cases Of ACC Suggesting That Hormonal Dependence Of ACC Like Carcinoma Breast (1). ACC Of Parotid Gland Typically Presents As Slow Growing Tumor In The Parotid Region With Mild Pain And Facial Nerve Palsy In Some Previous Reported Cases. Clinical Presentation Of ACC With Metastatic Lymph Node Or Metastatic Deposits Is Unusual Finding (4).

On Histopathological Examination, Usual Presentation On Gross Examination As Solitary Lesion, Usually Well Encapsulated With Soft In Consistency And Cut Surface Shows Greyish-White In Appearance. Microscopically, The Commonest Presentation Of Tumor Is Serous Acinar Cell Differentiation With Several Other Subtypes Include Intercalated Ductal, Acinar, Clear, Vacuolated, Microcystic, As In Present Case, Non Specific Glandular And Solid Lobular, Follicular And Papillary-Cystic Growth Patterns (8). Tumor Cells Are Round Or Pleiomorphic With Moderate To Abundant Lightly Basophilic Cytoplasm With Presence Of Fine Or Course Granules With Few Have Cytoplasmic Vacuolation Or Clearing. The Nuclei Are Eccentrically Placed With Inconspicuous Nucleoli (2). Few Microscopic Features Are Frequently Correlated With Aggressive Nature Include Frequent Mitotic Figures Per High Power Field, Tumor Necrosis, Per neural Or Lymphovascular Invasion, Marked Pleomorphism Of The Tumor Cells, Infiltration Into Surrounding Structures, Presence Of Stromal Hyalinization Changes And Those Cases Which Shows Dedifferentiation Of The Tumor From A Lower Grade To A Higher Grade Of Malignancy (1). Some Differential Diagnosis Always Kept Include Oncocytomas, Warthin’s Tumor, Clear Cell Carcinomas, And Mucoepidermoid Carcinomas (2). FNAC Examination Had Been Well Established Diagnostic Tool For Diagnosis Of Salivary Gland Lesions As It Provides Useful Information Regarding The Diagnosis And Further Treatment Of These
Tumors. FNAC Findings Are Usually Characterized By Abundant Acinar Differentiated Tumor Cells Present Within A Clean Background. Cells Mainly Arranged In Clusters Or With Microacinar Grouping. Tumor Cells Are Mild To Moderately Pleomorphic With Abundant Fragile, Vacuolated Cytoplasm And Nucleus Showing Mild To Moderate Anisokaryosis With Bland Chromatin. ACC Of Salivary Gland Is Commonest Cause Of False Negative Interpretation Due To Absence Of Morphological Characteristic Features Of Malignancy Such As Absence Of Markedly Pleomorphic Tumor Cells, Absence Of Tumor Necrosis, And Absence Of High Mitotic Or Proliferative Activity (9).

The Utility Of Radiological Examinations Are For Pre-Surgical Evaluation And Management Of The Tumor. USG Is Useful For Evaluation Of Tumor Size, Its Location, And The Nature Of The Growth. CT scans Usually Demonstrates Contrast Enhancement of the Growth and Useful Tool for Evaluation of Size of Growth, Extension of the Tumor, Relationship of Tumor to Facial Nerve and Other Structures, And Presence of Distant Metastatic Deposits (10).

The Management Of ACC Consists Of Complete Surgical Removal Of The Growth, By Total Or Subtotal Parotidectomy, And Postoperative Radiation Therapy Which Might Be Useful For Recurrent Lesion And Undifferentiated Or Dedifferentiated Cases Of ACC, Positive Surgical Margins, And Advanced Tumor Stage With Cervical Lymph Node Or Metastatic Spread Of The Growth (4). ACC Usually Considered As Chemoresistant Tumor, Which Is Probably Due To Its Slow Rate Of Metabolism (3). Few Prognostic Factors Of ACC Are Age Of The Patient, Association Of Pain Or Tenderness, Sex, Race, Previously Inadequate Surgical Removal Or Positive Surgical Margins, Advanced TNM Stage, And Invasion Of The Growth To The Base Of Skull (7).

4. Conclusion:
ACC Is Rare Malignant Tumor, Frequently Arising In Parotid Gland And Metastasis To The Intra-Salivary Gland Lymph Nodes Is Very Rare Finding In Young Male. Intra-Operative Frozen Section Might Be Helpful In The Diagnosis Because Of Low Specificity And Sensitivity Of CT Scan And FNAC Of ACC. The Histo-Morphological Examination Of Paraffin Embedded H/E Stained Section Is Still The Main Tool To Explain The Microscopic Findings Of ACC And To Classify ACC Of Parotid Gland. Surgery with Adjuvant Post-Operative Radiation Therapy Is Acceptable For Final Outcome of Disease. The Tumor Should Be Managed Uncompromisingly, Despite Of Its Slow Growing and Low Malignant Behaviour, Due To Its Metastatic Prospective Which Usually Confirmed By Distant Metastasis and Recurrence. Hence, These Tumors Require A Long-Term Follow-Up.

5. References:
6. Keane WM, Denny JC 3rd, Atkins JP Jr, Mebbrearty F. Acinic Cell Carcinoma of the


